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SURGICAL PRACTICE

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BRITISH SURGICAL PRACTICE

Under the General Editorship of

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HODGKIN'S DISEASE, OTHER RETICULOSES, RETICULO-SARCOMA AND MYELOMATOSIS

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GENERAL INTRODUCTION

Proliferation of reticulum

185.] The lymphoid, or lympho-reticular, tissue is subject to a number of disorders characterized by proliferation of reticulum with subsequent differentiation into one, or several, of the more adult cell types which normally spring from this multipotential mesenchymal ancestor. The initial clinical feature of these disorders is, usually, enlargement of lymph nodes, limited sometimes to those of one group; with the passage of time, however, new formations are found in all organs abundant in reticulum. They thus become, or may start as, systematized affections, and the proliferative process may result in a generalized enlargement of lymph nodes, liver and spleen, and an overgrowth of lymphoid tissue in the upper respiratory and alimentary tracts, in the skin and in the bone marrow. This is to instance but a few of the more commonly affected organs, for undifferentiated mesenchymal cells abound throughout the body and may undergo proliferation in any organ or tissue.

Commonly affected organs

The proliferative processes are of two types; the one, frankly neoplastic, may be called reticulosarcoma; the other, a progressive hyperplasia which lacks the microscopical criteria of tumour formation, is spoken of as reticulosis (Letterer, 1924). This concept of a "group of diseases of reticulum—in which proliferation is followed by differentiation into one or several of the possible cell progeny"—is due to Pullinger (1932) and, on this basis, Robb-Smith (1938) has elaborated a histological classification of these diseases.

PART I
HODGKIN'S DISEASE

1. DEFINITION

Hodgkin's disease, or lymphadenoma, is a reticulosis in which the proliferated reticulum becomes differentiated into collagen fibrils and myeloid cells. It differs from many other diseases of this group by starting as a local affection of lymph nodes and becoming generalized only in the later stages.

2. AETIOLOGY

Age and sex incidence

Hodgkin's disease is more common in the male than in the female; the onset is usually between the ages of 20 and 35 years, but has been described at all ages from 3 to 80 years. Geography, race and social status do not appear to affect its incidence.

Causation

The cause of the disease is unknown. During the past 50 years, attempts have been made to inculpate various infective agents—diphtheroids, mycobacteria, spirochaetes, yeasts and a virus. The role of all these agents, however, has been discredited. There is no agreement even concerning the nature of the disease; some regard it as a neoplasm, many as a granuloma. The view taken here, that it is a systematized proliferation of reticulum of specific type but unknown aetiology, avoids begging the question of causation.

Nature

3. PATHOLOGY

(1) Morbid anatomy

At first, the enlarged lymph nodes are of the consistency of soft rubber but, with the progress of the disease, they become increasingly firm, due to the fibrosis which comes to dominate the morbid process. In the early stages, the cut surface is pearl-pink but, with the increasing fibrosis, it takes on a whiter hue. Periadenitis is absent. In the spleen scattered islets of lymphadenomatous tissue contrast with the plum-coloured pulp; focal or diffuse formations of like tissue may be seen in the liver, lungs and kidneys; nodules are common in the bone marrow and lymphadenomatous lesions have been found in every organ of the body.

The lymph nodes

The spleen

Other organs

(2) Morbid histology

Microscopically, the features of the disease have been most extensively ... pro- ...)sule, ... e the node into irregular lobules of cellular proliferation. In these islands reticulum

The lymph node

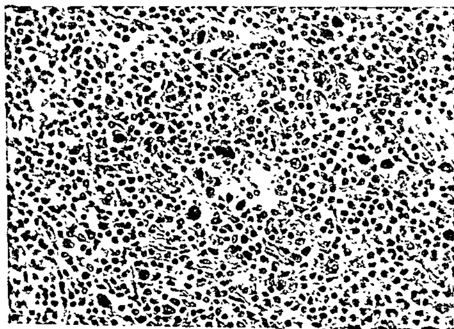


FIG 1 —Hodgkin's disease—cellular type with numerous giant cells and diffuse reticulum-cell hyperplasia throughout the medulla.

cells are notably increased, and their capricious division results in the formation of giant cells of various types. The most striking of these, the Sternberg-Reed giant cell with mirror image nuclei, has often been claimed as specific for Hodgkin's disease, but it may be seen in any reticulosis in which rapid irregular division of reticulum is in progress (Fig. 1) Accompanying the proliferation of reticulum cells is an increase in granulocytes; most frequently these are eosinophils and usually myelocytes, although neutrophils and

The Sternberg-Reed giant cell

basophils, and more mature forms of all three series, are not rare. Hyperplasia of fibroblasts and of cells producing reticulum is also evident, and there is commonly great increase of argyrophil fibrils (Fig. 2).

In the early stages, cellular proliferation often dominates the microscopical picture, fibrosis following later; lesions, however, are often found at different stages in various lymph nodes.

Other organs

The histological appearances in affected organs are similar to those in the

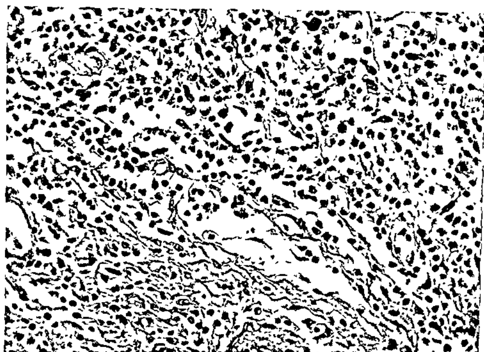


FIG 2.—Hodgkin's disease—sclerocellular type. There is proliferation of collagen.

lymph nodes, but the changes are more patchy and less fully systematized than in other reticuloses.

Histogenesis

Pullinger (1932) has shown that the histogenesis of Hodgkin's disease is best explained as a proliferation of reticulum with subsequent differentiation into fibroblasts, reticulin-forming cells and myeloid elements. It may be described as a fibro-myeloid medullary reticulosis (Robb-Smith, 1938).

4. CLINICAL PICTURE

(1) General

(a) The localized phase

The patient with Hodgkin's disease commonly comes under observation with enlargement of the lymph nodes of one superficial group; in 80 per cent of cases the nodes of one or more of the cervical groups are first affected. At this stage health is often unimpaired, although some loss of weight, indefinable malaise and pruritus may be noted. In the majority, radiography shows some enlargement of mediastinal lymph nodes. Irradiation causes the nodes to shrink and is followed by remission for from 6 to 9 months; other swellings then appear, perhaps in the same site, possibly elsewhere; a shorter remission follows the second course of radiotherapy.

(b) The generalized phase

Cachexia

Inexorably the disease spreads with the appearance of enlarged lymph nodes in most superficial groups and within the body cavities; the spleen and liver also become palpable. Before this, deterioration in general health can be noted; progressive loss of weight, anaemia and an earthy complexion are accompanied by fever and languor. Radiotherapy loses its effect; there may be effusions into the serous cavities and, after a course of from 2 to 7 years, febrile cachexia and death supervene.

(c) Variations in the course

Pel-Ebstein fever

This pattern is pursued with remarkable constancy, but variations are seen. The initial nodal swellings may be in some group other than the cervical; rarely, splenomegaly or a bone tumour is the presenting symptom. The course may be punctuated by the appearance of lesions in less common sites, such as the vertebral column, the skin or the meninges. The temperature chart may show the periodic pyrexia, familiar as Pel-Ebstein fever. The disease may pursue a more rapid or a more protracted course; some cases have terminated in less than 1 year and some have survived as long as 14 years. Instances of "acute Hodgkin's disease" ending fatally in a few weeks are usually examples of some other form of reticulosis.

(2) Lymph-node enlargement

Enlargement of lymph nodes

Enlargement of the nodes of one group is by far the most frequent initial symptom. The affected nodes are commonly those of a group in the neck, usually the lower deep cervical or the supraclavicular. It is said that the disease begins more commonly on the left than on the right side; however, in a series of 68 cases, which included those with enlarged nodes in many parts of the body, swellings were noted first on the left side of the neck in 33 per cent (Fig. 3), on the right in 35 per cent, and simultaneously on both sides in 10 per cent.



Fig. 3.—The cervical type of Hodgkin's disease: (a) before radiotherapy, and (b) after radiotherapy

It is unusual to see a patient with enlarged cervical lymph nodes in whom there is not already radiographic evidence of mediastinal disease. Of 53 such patients there were enlarged mediastinal or hilar shadows in 25, and in many of the others skiagrams were not taken at this early stage. It is true to say that in more than 50 per cent of cases there is disease of mediastinal, as well as of cervical, lymph nodes when the patient is first seen.

Initial enlargement of other lymph nodes

Other superficial groups are less frequently the primary seat of disease; typical figures are axillary 9 per cent, inguinal 1·5 per cent. Septic infection of the hand or arm not infrequently precedes axillary swellings. In 4·5 per cent of

(a) Characteristics of the enlarged lymph nodes

The degree of enlargement is variable; commonly 2 or 3 nodes are found, of diameter varying from 1 to 5 centimetres; at times large tumours are formed. The individual nodes are discrete, insensitive and mobile; in consistence they have a firm elasticity, becoming harder as the disease progresses and more fibrous tissue is elaborated.

In the later stages the nodes of most of the superficial groups become palpably enlarged.

(b) Splenomegaly

Splenomegaly

Splenomegaly is present at some time in more than two-thirds of all cases. It is commonly a late feature, but in about 3 per cent of cases it brings the patient under observation. It is usually moderate in degree, but, rarely, huge splenic tumours are seen. In some instances the splenomegaly is responsible for a secondary haemolytic anaemia or thrombocytopenic purpura.

(c) Hepatic enlargement

Jaundice and ascites

Hepatic enlargement of minor grade is almost constant in the later stages of the disease; great enlargement is uncommon. Jaundice is not rare in the closing phases and is usually due to masses in the porta hepatis. During this stage ascites may occur.

Intrathoracic lymph nodes

Mediastinal lymph nodes are affected in the majority of cases; in about 5 per cent their enlargement first brings the patient under observation but in more than 50 per cent there is radiographic evidence of their affection from the start of the illness. Although they often form large tumours, pressure phenomena are rare; obstruction of the superior vena cava is seldom seen and, when present, indicates an infiltrative lesion of the mediastinum.

Pulmonary lesions and serous effusions

In the later stages, lesions of the pulmonary parenchyma are common, either as multiple deposits throughout the lungs or as massive infiltrations.

Pleural and pericardial effusions may occur in the generalized phase of the disease. The fluid has the characteristics of a serous lymphocytic exudate.

(3) Skeleton

The osseous lesions of Hodgkin's disease have a particular interest for the surgeon. They usually appear late in the disease and give rise to symptoms in about 20 per cent of cases although this is but half the frequency as found at necropsy. The bones most commonly affected are the vertebrae, the sternum, the pelvis, the ribs, the skull and the upper ends of the femora and humeri.

Vertebral lesions

Pain also draws attention to bone deposits in most cases. With vertebral disease it may be a localized ache or have a radicular distribution. The vertebral column is usually invaded by direct extension from a mass of prevertebral lymphadenomatous tissue from which extensions may enter the extradural space; thus, compression of the spinal cord often occurs without vertebral collapse and sometimes without radiographic evidence of vertebral disease; in other cases gibbus is obvious.

Other bones

A sternal tumour is sometimes the presenting symptom; it is usually associated with mediastinal disease. Nodules may develop in the skull or ribs and occasionally deposits in the long bones lead to spontaneous fracture.

(4) Skin

Pruritus, unassociated with eruption, is a frequent early symptom; it occurs at some time in 25 per cent of cases. Pruriginous rashes are common and herpes zoster complicates the course in about 10 per cent of cases. The specific skin infiltrations are far less common.

Pruritus and herpes zoster

(5) Other lesions

Other organs are less commonly affected, but it is probable that every organ and tissue has, at some time, been the seat of lymphadenomatous deposit.

5. SPECIAL AIDS TO DIAGNOSIS

(1) Biopsy

Excision of a diseased lymph node for histological examination is the only method by which the diagnosis can be established. It is an essential step in every case.

(2) Lymph-node puncture

Lymph-node puncture has been advocated, but its results are uncertain, for smears made from the aspirated material do not disclose the alterations in the structure of the node on which microscopical diagnosis is based.

(3) Blood count

There is no characteristic change in the blood picture in Hodgkin's disease. In the early phases the blood count is usually normal; later, orthochromic or hypochromic anaemia is constant, often accompanied by a neutrophilia of 15,000–20,000 per cubic millimetre. Despite frequent statements to the contrary, eosinophilia is uncommon, although a few examples of extreme eosinophil leucocytosis have been recorded. A symptomatic haemolytic anaemia has sometimes been noted in the later stages. Bone marrow, obtained by sternal puncture, shows no constant or specific change.

Anaemia

The leucocytes

Sternal puncture

Blood and bone-marrow examinations are of value in excluding such related disorders as lymphatic leukaemia.

(4) Radiology

The chief importance of radiology is in determining the extent of the disease. A skiagram of the chest is essential, for in the majority of patients presenting with enlarged cervical or axillary lymph nodes there is also mediastinal disease.

(a) *The chest*

The radiographic changes are commonly those of mediastinal nodal enlargement, which may be sufficient to form a large tumour. In most cases the borders of the shadow are sharp-cut and its outlines rounded; it is sometimes possible to distinguish the margins of several individual nodes (Fig. 4). In the later stages parenchymatous disease is often present: irregular opacities may be scattered throughout the lung fields; a complete lobe may appear consolidated; or infiltrations may spread out from the hila. Pleural and pericardial effusions are not uncommon.

Parenchymatous disease

(b) *The skeleton*

The osseous deposits produce changes varying with the affected bone (Fig. 5). In the sternum, pelvis, ribs and femur they usually appear as rarefied

areas. The vertebral lesions are often osteoplastic, but, less commonly, they cause collapse of a vertebral body. In both cases an associated mass of lymphadenomatous tissue may cause a shadow similar to that of the paravertebral abscess of Pott's disease. When combined with vertebral collapse, differential diagnosis may be difficult. Sometimes periosteal infiltrations lead to erosion, often with new bone formation.

Periosteal infiltrations leading to erosion

(5) Specific tests

(a) *Erythrocyte sedimentation rate*

The erythrocyte sedimentation rate is normal at the onset but in the later stages is greatly accelerated. It has little value in differential diagnosis.

(b) *Mantoux test*

The tuberculin test may sometimes help to distinguish Hodgkin's disease from tuberculous lymphadenitis; it is usually negative, even with high concentrations, in sarcoidosis.

(c) *Wassermann reaction*

The Wassermann reaction is found to be positive in about 1 per cent of patients with Hodgkin's disease, but it is seldom required to exclude the lymphadenitis of secondary syphilis.

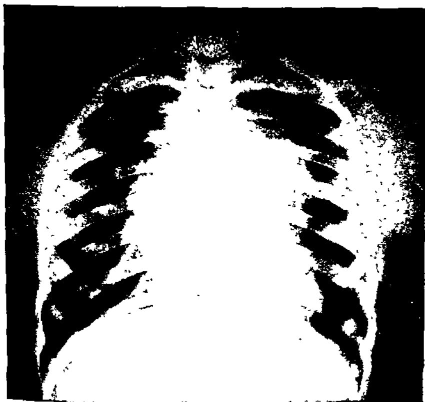


(a)

stinal lymph nodes in the generalized phase of the disease.



(b)



(c)

(d) Plasma proteins

The plasma proteins show no constant alteration in Hodgkin's disease; a rise in the globulin content of the plasma with enlargement of lymph nodes favours the diagnosis of sarcoidosis.

6. DIFFERENTIAL DIAGNOSIS

Other reticuloses and reticulo-sarcoma

The common cervical type of Hodgkin's disease requires differentiation from all other causes of enlargement of lymph nodes in the neck. Such causes fall into three groups. First, other reticuloses and reticulosarcoma, from which distinction can usually be made only by histological study; lymphatic leukaemia can be excluded by blood and bone-marrow examinations. Secondly, metastatic carcinoma, in which the nodes are usually hard and fixed, and in which investigation will divulge a primary tumour perhaps of the bronchus, nasopharynx or stomach. Thirdly, inflammatory enlargement: pyogenic lymphadenitis seldom offers difficulties, but furunculosis and sarcoidosis may require biopsy to ensure differentiation (Plate I (b)). In patients returning from endemic areas trypanosomiasis should not be forgotten. Lateral aberrant thyroid masses have sometimes been mistaken for enlarged lymph nodes.

Lymphadenitis



(a (i))



(a (ii))



(a (iii))



(a (iv))

FIG 5.—Skiagrams of osseous lesions in Hodgkin's disease. (a) Vertebral lesions: (i) osteolytic lesion with collapse of fifth lumbar vertebra, (ii) osteolytic lesion with collapse of eighth dorsal vertebra Antero-posterior view, showing also the paraverte-

(e) Osteolytic lesion of upper end of right humerus.


(b (i))


(b (ii))

(c)


(d)



(e)

7. DIAGNOSIS

Trousseau (1877) declared that a diagnosis of Hodgkin's disease was a sentence of death. His observation remains true today. There are occasional cases in which recovery appears to have taken place but over 50 per cent of patients die within 3 years, *and after 5 years not more than 10 per cent* are still living.

8. TREATMENT

(1) General

Increased liability to infection

The patient with Hodgkin's disease should be encouraged to lead a normal life for as long as possible. His increased liability to infection should be borne in mind and common colds and other minor disorders should be treated more seriously than in the otherwise healthy man.

(2) Radiotherapy

The "bath" method of irradiation

The various forms of "radical" treatment aim at destruction of the pathological tissue. Radiotherapy is the most effective measure we possess at present. The technical methods used are several: some workers favour small doses applied locally, sufficient only to cause shrinkage of the enlarged nodes; others prefer local application of larger doses, hoping thereby not only to destroy the local mass but to prevent recurrence in the same site; yet others recommend the "bath" method, whereby the whole thorax, abdomen or trunk is irradiated simultaneously. The last procedure requires careful haematological control and is not without danger.

(3) Chemotherapy

Nitrogen mustards

Chemotherapy in the form of administration of arsenic has been used for many years; its results, however, are indifferent. More recently, compounds of the nitrogen-mustard series, particularly methyl-bis (β-chloroethyl)amine hydrochloride, have been found to cause temporary remission in some cases. This substance is given intravenously in doses of 0·1 milligram per kilogram of body-weight for 4 consecutive days. Its place in treatment is not yet established (Rhoads, 1946; Jacobson and others, 1946).

9. INDICATIONS FOR SURGICAL INTERVENTION

(1) Diagnostic biopsy

Choice of site for biopsy

Diagnostic biopsy is the main indication for surgery in Hodgkin's disease. When numerous lymph nodes are enlarged, one from the lower deep cervical or supraclavicular group is the best choice, for the microscopic structure of upper deep cervical, inguinal and axillary nodes is often distorted by fibrosis, residual from a past lymphadenitis.

(2) Radical excision

Total extirpation of the diseased nodes has been decried as a therapeutic [illegible]

radiotherapy gives better results than do other methods (Baker and Mann, 1939). Suitable cases are few, but this method should be considered in the type of case in which the disease is limited to the upper deep cervical group of nodes on one side.

(3) Splenectomy

Splenectomy is occasionally indicated in Hodgkin's disease. Rarely, patients are seen whose disability is largely due to chronic haemolytic anaemia or to thrombocytopenic purpura secondary to a lymphadenomatous splenomegaly; in such cases, splenectomy relieves the symptom and may provide 1 or 2 years of comfortable existence. When there is pain due to a greatly enlarged spleen without evidence of rapidly advancing disease elsewhere, splenectomy may considerably increase the patient's comfort.

Vertebral collapse and pathological fracture require management on the usual orthopaedic lines.

PART II
OTHER RETICULOSES

1. DEFINITION

The general concept of the reticuloses as proliferative conditions of reticulum, with later differentiation, has already been discussed.

2. CLASSIFICATION

The classification of the reticuloses, other than Hodgkin's disease, employed here is that of Robb-Smith (1938), and is histological and descriptive. It is based on the changes found in the diseased lymph nodes. Proliferation may start in the follicles, the medulla or the sinuses: the reticuloses resulting are thus follicular, medullary or sinus—these descriptions being qualified by a

Lymph-node changes

term denoting the dominant cell type. Many of these disorders are familiar under different names, but there is no other nomenclature which so clearly shows the essential unity of the group. The number of histological types is considerable; they include the lipoidoses, which are storage reticuloses, as well as the leukaemias. It is impossible to discuss all the varieties here; those that require comment are:

1. Lymphoid follicular reticulosis (follicular lymphoblastoma of Brill, Baehr and Rosenthal, 1925).
2. Lymphoid medullary reticulosis (lymphoid leucosis or leukaemia).
3. Lympho-reticular medullary reticulosis (Hodgkin's paragranuloma, Jackson and Parker, 1944).
4. Histiocytic medullary reticulosis (Scott and Robb-Smith, 1939).

3. LYMPHOID FOLLICULAR RETICULOSIS

(1) Incidence

Lymphoid follicular reticulosis has been recognized since its description by Brill, Baehr and Rosenthal (1925). It is not rare, being seen about once for every 6 cases of Hodgkin's disease. Its main incidence is between the ages of 30 and 50 years, and both sexes are affected equally.

(2) Pathology

Histological changes

Pathologically there is generalized enlargement of lymph nodes, splenomegaly and hepatomegaly; serous effusions are common. All diseased tissues reproduce the histological changes seen in the lymph nodes (*see* Plate I (a)). The follicles are increased in size and number, compressing the pulp; each hyperplastic follicle presents a central area of lymphoblasts, surrounded by a rim of small lymphocytes; intermediate grades of lymphoid cells, curiously, are lacking (Figs. 6, 7, 8 and 9). A proportion of these cases terminate in sarcomatous change.

(3) Clinical picture

Enlargement of lymph nodes and spleen

The common clinical picture is one of generalized enlargement of lymph nodes with splenomegaly and little disturbance of the general health. Occasionally the patient comes under observation with massive splenomegaly or predominant enlargement of one group of lymph nodes. In many cases effusions into serous cavities occur early, and such an event does not indicate, as in Hodgkin's disease, that the final phase of the disorder has been reached. Chylous effusions are not rare, and deposits in the orbit and breast have been recorded.

(4) Prognosis

The course is prolonged and the masses respond readily to radiotherapy. The majority of patients live for 5 years and survival for 10 or more years is not rare. In a considerable proportion of cases the end comes with malignant degeneration in one group of nodes, in which local invasive phenomena become evident.

(5) Diagnosis

Diagnosis can be made with certainty only by biopsy of an enlarged node, and this should always be undertaken. In some cases sarcomatous change has been accompanied by a notable lymphocytosis of the peripheral blood.



(*a*)



(*b*)

(*a*).—Lymphoid follicular reticulosis. Section of lymph node, showing greatly enlarged and hyperplastic follicles. (*b*) —Boeck's sarcoid. Section of lymph node, showing typical sarcoid lesions with giant-cell formation.

PLATE I



FIG. 6.—Lymphoid follicular reticulosis: low-power view, showing hyperplastic follicles.

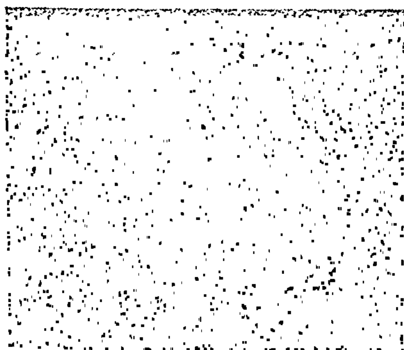


FIG. 7.—Lymphoid follicular reticulosis: high-power view of an enlarged and hyperplastic follicle

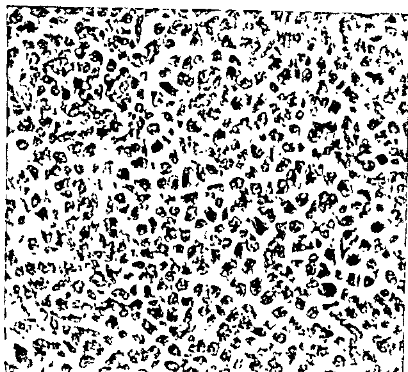


FIG 8.—Lymphoid follicular reticulosis: high-power view.

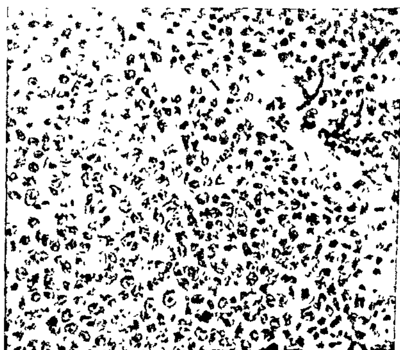


FIG 9.—Lymphoid follicular reticulosis undergoing malignant transformation; many lymphoblasts are evident.

(6) Treatment

Deferment of irradiation

Radiotherapy is the only satisfactory method of reducing the local masses, which are remarkably radio-sensitive. It is advisable to defer irradiation until

it is urgently demanded by symptoms, for there is some evidence that it may hasten the neoplastic metamorphosis.

Indications for Surgery

Indications for surgery are few, as the disorder is usually generalized when first seen; the only constant one is biopsy. In some cases random splenectomy for unexplained splenic tumour reveals that the organ is affected by this disease. When the diagnosis has been established but a large splenomegaly is causing pain, palliative splenectomy may be a reasonable procedure.

4. LYMPHOID MEDULLARY RETICULOSIS

(1) Pathology

This term is no more than a description of the histological changes in the lymph nodes in lymphoid leukaemia; all sign of the normal structure is obliterated by a diffuse proliferation of small lymphocytes. *Proliferation of small lymphocytes*

(2) Surgical significance

The disorder has some importance for the surgeon because, although it is commonly manifested as a generalized disease, local infiltrations may lead the patient to seek surgical advice. One lymph-node group may show predominant enlargement, or great splenomegaly may be the only sign of disease. In the more acute forms in childhood a subperiosteal infiltration sometimes occasions pain of sufficient severity to raise the suspicion of osteomyelitis. Proliferation in the pharyngeal lymphoid tissue may cause deafness or may result in indolent painful ulceration. *Local infiltrations*

(3) Diagnosis

Blood counts will usually show a lymphocytosis, but in some instances sternal puncture and even microscopical examination of an excised lymph node will be necessary to make diagnosis secure. *Lymphocytosis*

5. LYMPHO-RETICULAR MEDULLARY RETICULOSIS

(1) Incidence

This disease has been described by Jackson and Parker (1944) as Hodgkin's paragranuloma, with the assertion that it may be the forerunner of true lymphadenoma. This contention is not endorsed by others and the condition is considered here as a distinct type of reticulosis It affects an older age-group (30–40 years) than does Hodgkin's disease and the sex incidence is about equal. *Age-group and sex incidence*

(2) Pathology

Microscopically the lymph node shows a diffuse infiltration of small lymphocytes throughout the medulla, but scattered amongst them are variable numbers of reticulum cells; traces of the normal structure are sometimes still apparent.

(3) Clinical picture

Lympho-reticular medullary reticulosis is essentially a localized disorder in the early stages and, in most instances, a mass in the neck brings the patient under observation. There is no disturbance of general health or other sign of disease, and 5 or more years may elapse before a lesion appears elsewhere. This lesion is often at some site far removed from the lymph nodes initially *Localized disorder*

Paraplegia relatively common

affected; paraplegia is a relatively common symptom. It is the most benign of the reticuloses, and survival for as long as 30 years has been reported.

(4) Treatment

The local masses respond well to irradiation, but it would seem that this condition, above all, is suitable for radical surgical excision of the affected nodes followed by radiotherapy; only its wider recognition will permit assessment of the value of this treatment.

6. HISTIOCYTIC MEDULLARY RETICULOSIS

(1) Incidence

This rare disorder was described by Scott and Robb-Smith in 1939. Several cases have been reported since. It affects young and middle-aged adults.

(2) Pathology

The lymph node, throughout the medulla, shows proliferation of actively phagocytic cells containing erythrocytes and cellular debris (Fig. 10).

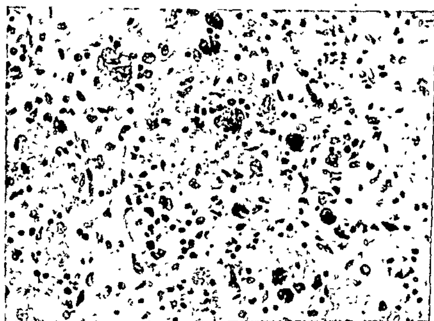


FIG. 10—Histiocytic medullary reticulosis—showing hyperplasia of large phagocytic cells throughout the medulla; some of these cells contain erythrocytes.

(3) Clinical picture

Rapid course

The course of the disease is rapid, with fever, emaciation, increas anaemia and a profound leucopenia. Soft lymph nodes are usually palp in all groups and the spleen is enlarged, sometimes greatly.

(4) Surgical significance

Response to splenectomy

The surgical interest of the condition lies in the response to splenecto In several cases this operation has been followed by a remission lasting 3 more months; without splenectomy, deterioration is rapid and survi seldom exceeds 10 weeks. The diagnosis must be confirmed by lymph-n biopsy before splenectomy is undertaken.

PART III
RETICULOSARCOMA

1. DEFINITION

A reticulosarcoma is a neoplastic proliferation of mesenchymal cells or of their progeny, leading to stromal destruction as well as infiltration.

2. AETIOLOGY

Reticulosarcomas are seen throughout the span of life, but the commoner varieties usually affect adults of middle age. Its incidence in the two sexes is equal. A proportion arise in established cases of reticulosis, particularly of the lymphoid follicular type. No other aetiological factors are known.

Age and sex incidence

3. PATHOLOGY

(1) Morbid anatomy

Reticulosarcomas may occur as localized tumours with spread to regional lymph nodes and with blood-borne metastases, or they may be systematized throughout the lympho-reticular tissue, even when the disease starts as a

localized process systematization usually supervenes with the passage of time, unless the local tumour proves lethal for some mechanical reason. The

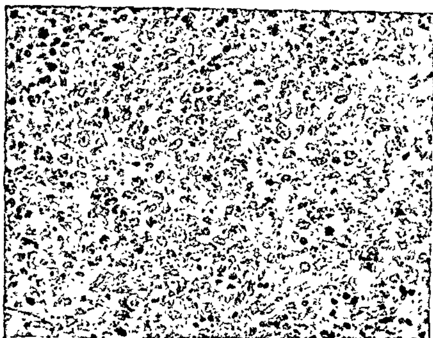


FIG 11.—Syncytial reticulosarcoma—showing undifferentiated syncytial structure.

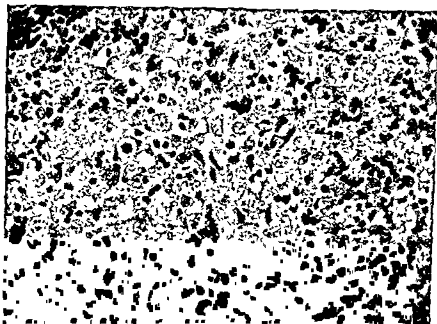


FIG 12.—Dictyosyncytial reticulosarcoma This is somewhat more differentiated than is the reticulosarcoma of Fig. 11, having a syncytial structure together with occasional dictyocytes.

characters of the tumours vary with their location; in general, they are large, firm, irregularly rounded masses; the cut surface is white, and haemorrhages and areas of necrosis are common.

(2) Morbid histology

A number of histological varieties of reticulosarcoma can be recognized, but their biological behaviour does not appear to depend very closely on their cytological character. A classification, on a cytological basis, was suggested by Géry and Bablet in 1935, and Robb-Smith's modification of this (1938) is followed here.

Biological behaviour in relation to cytology

(3) Histological classification

These tumours may be divided into undifferentiated, histioid and haemic varieties. The undifferentiated or syncytial reticulosarcoma consists of synplasmic sheets of pale staining cells with vesicular nuclei; in some a stroma reaction produces a trabecular pattern (Fig. 11). The histioid or dictyocytic reticulosarcoma is composed of polygonal cells with darkly staining nuclei and a profuse intercellular reticulin meshwork. Intermediate between these is the dictyosyncytial variety (Fig. 12). These three tumours are rare.

Undifferentiated histioid and haemic varieties

(*a*) *Lymphoid reticulosarcomas*

By far the commonest lymphoid reticulosarcomas are those in which there is differentiation into blood cells: the lymphoblastic and lymphocytic reticulosarcomas (lymphosarcoma). Lymphoblastic reticulosarcomatous change may supervene in lymphoid follicular reticulosis when the follicular pattern of the original disorder is, to some extent, preserved Into this group fall also multiple myelomatosis (plasma-cell reticulosarcoma) and chloroma (myeloblastic reticulosarcoma).

(*b*) *Polymorphic reticulosarcomas*

Finally, a polymorphic variety occurs in which both haemic and histioid elements are represented; this has been described as "Hodgkin's sarcoma" or "malignant Hodgkin's disease". Its microscopical resemblance to true lymphadenoma may be remarkable and it has often been held to develop in lymphadenomatous tissue, but the evidence for this is unconvincing.

"Hodgkin's sarcoma"

4. CLINICAL PICTURE

(1) Generalized type

One of the commonest clinical guises assumed by reticulosarcoma is that of generalized enlargement of lymph nodes with spleno-hepatomegaly. Enlargement of one group of nodes may predominate, but often all are only of moderate size. The general health is little impaired in the early stages. The initial response to irradiation is often satisfactory, but fresh masses appear rapidly and cachexia soon becomes evident. Diffuse infiltration of the bone marrow leads to anaemia and deposits may form in the skin and the upper respiratory and alimentary tracts. The course is seldom prolonged above 2 years. It is the lymphoblastic reticulosarcoma which most frequently leads to this clinical picture.

Course

(2) Localized types

Although it is the rule for the less common varieties to start in this manner, the lymphoblastic and lymphocytic reticulosarcomas form the highest proportion of these localized tumours, for they greatly outnumber all other histological types.

(a) Local lymph-node tumours

The initial mass may arise in one group of lymph nodes and, although the cervical groups are the commonest sites, the number of cases presenting with an inguinal tumour is considerable. When the mediastinal nodes are first affected lymphoblastic reticulosarcoma may be accompanied by the blood picture of lymphoid leukaemia; this combination has long been known as Sternberg's leucosarcoma.

Sternberg's leucosarcoma

(b) Leucosarcoma

Many cases of "retroperitoneal sarcoma" are reticulosarcomas arising in retroperitoneal lymph nodes; on occasion, haemorrhage into one of these tumours may lead to a clinical picture simulating that of an acute abdominal catastrophe, or fever and leucocytosis may suggest perinephric abscess.

(c) Bone tumours

Myelomatosis

Myelomatosis (plasma-cell reticulosarcoma) is the only variety commonly presenting as a tumour of bone. It is considered in detail in Part IV. Two clinical types occur: a diffuse infiltration of the bone marrow, particularly of the ribs, vertebrae and skull, and a similar infiltration combined with local tumours of these bones and sometimes leading to spontaneous fracture or paraplegia. Rare instances of solitary plasma-cell reticulosarcoma have been recorded but in the great majority systematization follows. Chloroma (myeloblastic reticulosarcoma) commonly occurs in children and gives rise to tumours beneath the periosteum of the skull and orbit; it is accompanied by the blood changes typical of myeloblastic leukaemia. The histogenesis of Ewing's tumour of bone is undecided but there is evidence that a proportion, at least, are trabecular undifferentiated reticulosarcomas.

Chloroma

Ewing's tumour

(d) Alimentary tract

The alimentary tract is not a rare site for reticulosarcoma. The stomach is sometimes primarily affected. Lymphoblastic, lymphocytic and polymorphic types may also occur in the ileum, caecum, ascending colon, and, less frequently, the jejunum They are probably the commonest tumours of the small bowel in which they lead to chronic obstruction often with intussusception. In some cases a syndrome closely resembling that of Addison's disease has been noted. In the later stages of generalized lymphoblastic reticulosarcoma multiple rectal tumours with obstruction or paradoxical diarrhoea may occur.

Tumours of the small bowel

(e) Nasopharyngeal tumours

The lymphoid tissues of the upper respiratory tract and of the tonsils are common primary sites for these tumours. They are usually of the lymphoid type and systematization is late.

5. SPECIAL AIDS TO DIAGNOSIS

(1) Diagnostic biopsy

Biopsy of a diseased lymph node or of the tumour itself is the only way of making a certain diagnosis. It should never be omitted.

(2) Blood count

Normocytic ...ia

In the later stages anaemia is constant; it is normocytic unless there has been loss of blood. The leucocyte count is variable: in the terminal phase of the lymphoid reticulosarcomas a leukaemoid blood picture may occur, but it

is usual for the lymphocytes to show a pleomorphism rare in lymphoid leukaemia; chloroma (myeloblastic reticulosarcoma) is accompanied by the blood changes characteristic of myeloblastic leukaemia, and myelomatosis (plasma-cell reticulosarcoma) occasionally by those characteristic of plasma-cell leukaemia.

(3) Sternal puncture

Sternal puncture is of the greatest value in the diagnosis of myelomatosis; even when the tumour is solitary, films of the sternal bone marrow often show a large percentage of the characteristic atypical plasma cells. In the lymphoid reticulosarcomas sternal puncture will often demonstrate a lymphoid transformation of the bone marrow, and in chloroma, films show 80–95 per cent of the nucleated elements to be myeloblasts.

Lymphoid transformation of bone marrow

(4) Radiology

Radiology can demonstrate the location of the various tumours, whether mediastinal, in the alimentary tract, or skeletal, but it cannot reveal their pathological nature, except perhaps in the last instance. The radiographic changes of these tumours of bone are described in Part IV.

(5) Other tests

The urine, in 60 per cent of patients with myelomatosis, contains Bence-Jones's protein, a great increase in the plasma globulin is common, and a "false positive" Wassermann reaction may occur.

Bence-Jones's protein

6. DIFFERENTIAL DIAGNOSIS

The differential diagnosis of reticulosarcoma varies with the site of the primary tumour. The other diseases causing enlargement of lymph nodes require exclusion; this can be done only with the aid of the microscope. In this regard two tumours, which sometimes confound the histologist, deserve mention: the nasopharyngeal lympho-epithelioma and the anaplastic carcinoma the metastases of which may resemble the syncytial reticulosarcoma. A detailed consideration of the differential diagnosis would include discussion of all other tumours likely to arise in the same sites as do the reticulosarcomas.

Variation with site of primary tumour

7. PROGNOSIS

Reticulosarcoma is a fatal disease. It is generally believed that the course of the undifferentiated types is more rapid than that of the lymphoid tumours, but the evidence for this is slender. The tumour may prove rapidly lethal by virtue of its position, for instance when it arises in the small intestine, or it may become systematized and result in cachexia terminating in death. The average period of survival does not exceed 2 years.

Period of survival

8. TREATMENT

(1) Radiotherapy

Radiotherapy, at present, is virtually the only treatment employed. In certain cases, particularly those of localized tumours of lymph nodes or of the nasopharynx, the initial result is satisfactory, but, in almost all, recurrence at the same site or systematization follows after a period, and the disease finally becomes radio-resistant. It is not rare to find initial response to

radiotherapy in a primary tumour, only to be followed in 2 or 3 months by rapidly fatal systematization. When there is severe anaemia and the process is acute, as in chloroma, irradiation is ineffective and dangerous.

Contra-indications

(2) Chemotherapy

Chemotherapy, including the use of nitrogen mustards, has not proved of value. Snapper and Schneid (1946) have claimed that a low-protein diet and injections of diamidinostilbene will cause some recession and will relieve the pain in myelomatosis.

(3) Surgery

Evidence is accumulating that, when possible, radical surgical excision followed by irradiation is more effective in prolonging life than is irradiation alone. There are relatively few suitable cases. In the rare instances of reticulosarcoma localized to the stomach patients may remain well for many years after gastrectomy. In reticulosarcoma of the small intestine the disease is almost always widespread within the abdominal cavity when the diagnosis is made.

9. INDICATIONS FOR SURGICAL INTERVENTION

(1) Diagnostic biopsy

Biopsy, either of a lymph node or of the tumour itself, is the chief indication for surgery. Diagnosis is seldom possible without the aid of the histologist.

(2) Radical surgery

When a local tumour exists, radical extirpation, as has been said, appears to give better results than does radiotherapy alone. It is likely that when removal of all diseased tissue is possible, as in the case of a superficial nodal mass, it should be recommended. Reticulosarcoma limited to the stomach is an indication for gastrectomy.

Indication for gastrectomy

In reticulosarcoma of the ileum, obstruction may indicate a palliative entero-enterostomy; resection is seldom, if ever, feasible.

PART IV
MYELOMATOSIS

1. DEFINITION AND AETIOLOGY

Myelomatosis, or Kahler's disease, is a neoplastic disease of bone marrow which must be regarded as a reticulosarcoma, systematized to a limited degree. The cell type is usually the plasma cell, but tumours occasionally develop from other bone-marrow elements. It is an uncommon disorder, rarely seen before the age of 40 years, and it occurs twice as frequently in men as in women. *Incidence*

2. MORBID ANATOMY

Multiple tumours, arising almost exclusively in actively haemopoietic bone marrow, characterize the disease; rarely, similar proliferations are found in the spleen, lymph nodes or liver. The individual tumours destroy the overlying bony cortex, and thus pathological fracture is common. On section they are soft dark-red, or grey-red, masses with areas of haemorrhage. *Morbid anatomy*

The microscopical appearance is of a sheet of ovoid cells, each about 10 μ in diameter, with an eccentric nucleus the chromatin of which is disposed in the "wheel-spoke" fashion typical of the plasma cell. The cytoplasm of the cells is basophilic and often vacuolated; a clear perinuclear zone is uncommon. Although these elements are often termed plasma cells, some prefer the description "myeloma cell", as it has no cytogenetic connotation. Rarely, the cell type in myelomatosis is the myeloblast, and still more rarely, the erythroblast. *Histology*

In some patients extensive obstruction to the renal tubules is found *post mortem*; this lesion may be sufficiently widespread to cause renal failure. The obstructing material appears to be Bence-Jones's protein, which is discussed in a later paragraph. *The renal lesions*

3. CLINICAL PICTURE

(1) "Solitary myeloma"

In some instances the patient seeks advice on account of a solitary tumour of bone, and neither clinical nor radiological examination may disclose other deposits. In almost all cases, the disorder will become systematized after the passage of 1 or 2 years.

(2) Pathological fractures

More frequently the widespread nature of the disease is obvious when the patient is first seen. Pain, local tumours and pathological fractures are the early features. The tumours are confined to the vertebral column, the skull, the bones of the thoracic cage, the pelvic bones and the upper parts of the humeri and femora. Thus local swellings may be noted on ribs, sternum or clavicles, and pathological fractures of ribs, clavicles and vertebral bodies are common. Pain depends on local tumour formation or on compression of nerve trunks. In many cases, ill-defined but severe backache is the first symptom; vertebral collapse may be associated with radicular pain and not rarely leads to compression of the spinal cord with paraplegia. Occasionally a myeloma of the basis cranii may compress cranial nerves. *Pain* *Paraplegia*

Occasionally, when there is a more diffuse infiltration of the skeleton, with less tendency to localized tumour formation, anaemia, or symptoms suggesting chronic nephritis, bring the patient under observation. In all cases, these *Anaemia*



Fig. 13 —Sternal-puncture material from patient with myelomatosis, showing a high proportion of myeloma cells of the plasma-cell type.

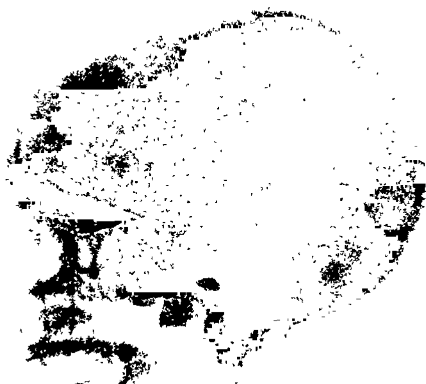


Fig. 14.—Skiagram of the skull in myelomatosis, showing areas of rarefaction in the vault and destruction of the sella turcica

general symptoms eventually make their appearance and the disease terminates with cachexia, anaemia, a tendency to spontaneous haemorrhage, and sometimes with amyloid disease. In some patients the progress[illegible] leads to death in uraemia.

The disorder is [illegible] and its durat[illegible] appear to be [illegible] [illegible]ment. [illegible] its course does not [illegible]

Course

4. SPECIAL AIDS TO DIAGNOSIS

(1) Sternal puncture

In myelomatosis there is proliferation of tumour cells throughout the bone marrow and they may be demonstrated in films of material obtained by sternal puncture. Stained by a Romanowsky method, the myeloma cell has a diameter of 15–25 μ with a circular eccentric nucleus some 6 μ in diameter. The nuclear chromatin is arranged in coarse clumps, but the "wheel-spoke" pattern is lacking: the cytoplasm is basophilic and often vacuolated. Giant and binucleate forms are common. Such cells can be found in the sternal marrow of most patients who appear to have a solitary myeloma, and sternal puncture is the most valuable single diagnostic method in this disease (Fig. 13).

(2) Radiology

The radiological changes are of two kinds: first, the appearance of multiple tumours of bone shown as rounded, punched-out areas of translucency, usually with sharp margins and occurring in the sites already mentioned. Secondly, a diffuse rarefaction without other change in the bony architecture. The first change is often best seen in the skull (Fig. 14); the second, in the vertebral column. In a proportion radiological changes are absent in the early stages.

(3) Blood count

An anaemia of normocytic type is invariable as the disease progresses; moderate leucopenia and thrombocytopenia are common. Rarely, the blood picture assumes the leuco-erythroblastic form, and yet more rarely a plasma-cell leukaemia occurs. Occasionally the clinical and radiological changes of myelomatosis are associated with the blood picture of myeloblastic leukaemia.

Anaemia

Blood picture of leukaemia

(4) Blood chemistry

A greatly raised value for the plasma proteins is present in about 60 per cent of cases. Figures exceeding 23 grammes per 100 millilitres have been recorded. The increase is in the globulin fraction, but the exact nature of the protein substance is unknown. It is presumed to be elaborated in the diseased bones. The serum-calcium level is often raised, but that of the inorganic phosphorus is normal. Retention of nitrogen is common in the later stages.

Plasma proteins

Serum calcium

The erythrocyte sedimentation rate is greatly increased and auto-agglutination of erythrocytes is common.

(5) Urine

In more than two-thirds of all patients with myelomatosis the abnormal protein first demonstrated by Bence-Jones can be found in the urine. It has

Bence-Jones's protein

the property of being precipitated at 50°–60° C. but of redissolving *when the* temperature is raised to boiling-point. Thus, when urine is heated, a flocculent white deposit is thrown down at about 60° C., redissolves when boiling-point is reached and reappears as the urine cools. The presence of Bence-Jones's protein in the urine is virtually pathognomonic of myelomatosis.

5. DIFFERENTIAL DIAGNOSIS

Diagnosis seldom offers difficulty when the possibility of myelomatosis is borne in mind, and radiology, sternal puncture and the assessment of bio-chemical changes will confirm or dispel the suspicion. Of other generalized diseases of the skeleton, carcinomatosis may present similarities and, when diffuse rarefaction is the only radiological change, senile osteoporosis and hyperparathyroidism require differentiation.

Carcinomatosis

Osteoporosis

6. TREATMENT

Irradiation

There is no curative treatment. Irradiation will relieve the pain of local tumours but obviously cannot be applied to the whole skeleton. Snapper and Schneid (1946) have shown that symptomatic improvement may follow the use of diamidinostilbene, by intravenous injection, in doses similar to those recommended in kala-azar. This therapy should be combined with a diet free of animal protein. Otherwise treatment is symptomatic.

Chemotherapy

7. INDICATIONS FOR SURGICAL INTERVENTION

Excision of tumours

There are few indications for surgery in myelomatosis. Biopsy of a tumour is rarely required. There are instances in which excision of an apparently solitary myeloma is justifiable; such a tumour resulting in pathological fracture of a clavicle is an example.

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[References to other titles are given under Hodgkin's Disease, other Reticuloses, Reticulosarcoma and Myelomatosis in the Index Volume. The subject is also dealt with under the heading of Hodgkin's Disease in the *British Encyclopaedia of Medical Practice* (1937), Vol. 6, p. 523]

HORMONES

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PAGE

1. INTRODUCTION

186.] The scope of this article is general rather than particular. Individual glands, such as the thyroid, adrenals and gonads; clinical conditions associated with hormone activity, such as pregnancy; disorders of endocrine function, such as pituitary disease, diabetes and menstrual disturbances; even certain hormones themselves, such as insulin, are dealt with in other articles in BRITISH SURGICAL PRACTICE. There is need, however, for an article setting out the principles of hormone action and endocrine therapy, the nature of the hormones and how they are administered, and the clinical methods of investigating endocrine function. These are the subjects discussed in the present article.

2. PRINCIPLES OF HORMONE ACTION

Definition

A hormone is a chemical substance, elaborated by an endocrine cell, to be secreted into the blood stream and to act upon specifically sensitive peripheral receptors.

(1) The endocrine cell

The hormone is elaborated in the secretory cells of the endocrine gland from raw materials brought to it in the blood stream. The thyroid cells, for instance, have an affinity for iodine and constitute the factory in which the hormone is manufactured from this essential component. Some gland cells, such as the chromophil cells of the anterior pituitary, contain granules composed of the hormone, which are therefore extruded when it is secreted into the blood stream, the cell then entering upon a phase of rest or secretory inactivity. Other endocrine cells contain pigments, such as carotene which gives the corpus luteum its yellow colour, or yield characteristic staining reactions due to the hormones they elaborate, as do the chromaffin cells of the adrenal medulla.

Phase of secretory inactivity

The rate of cellular activity varies from complete rest to a state of excessive function and the histology of the cell may consequently alter, as in the thyroid in which the resting cell is flattened against its basement membrane, whereas the overactive cell of thyrotoxicosis is columnar. The rate of activity is controlled by factors such as the supply of raw material, the demands of the tissues for the hormone and the influence of other glands. The trophic hormones of the pituitary, for instance, stimulate the cells of the thyroid, adrenal cortex and gonads. The various factors are indicated in the accompanying diagram (Fig. 15).

Variations in cell histology

Whether a single endocrine cell can secrete more than one hormone is still a matter of speculation. Such a supposition would explain the multiplicity of hormones of the anterior pituitary, which possesses only two types of secreting cell. On the other hand the cellular secretion may undergo various chemical changes in the circulation, leading to different hormonal effects, or

alternatively, one hormone may produce different effects depending upon the nature of the peripheral receptors which it stimulates.

There is little evidence, however, except perhaps in the case of adrenal cortical tumour, of an endocrine cell elaborating an abnormal or pathological type of hormone. For instance, there is no need to postulate that an abnormal

No pathological type of hormone

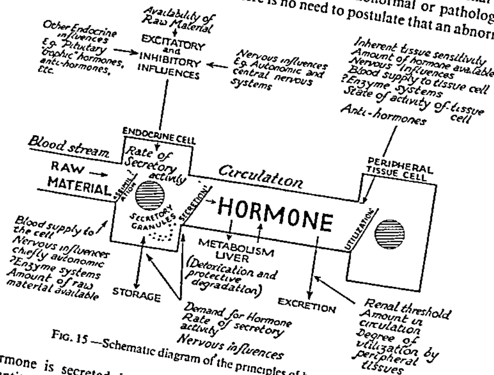


FIG. 15.—Schematic diagram of the principles of hormone action.

hormone is secreted in "thyrotoxicosis"; it is simply that an excessive quantity of normal thyroid hormone is manufactured.

In the case of the thyroid, facilities for storing the hormone are provided. In other endocrine organs the hormone may be stored in the cell itself.

(2) Circulation, metabolism and excretion of the hormone

The hormone generally is delivered constantly into the blood stream, though the amount may vary according to the rate of secretory activity of the cell and the demand for the hormone. Some believe that adrenaline may be an exception. In conditions of emergency there is no doubt that the circulation is flooded with the hormone, and that this inconstant supply is the principal mode of adrenaline action; it is probable, however, that a very slight continuous secretion from the adrenal medulla also occurs.

Constant supply to blood stream

Hormones such as insulin, adrenaline, pituitrin and thyroid hormone probably reach their peripheral target in the same chemical form as that in which they left the secreting cell (Fig. 16). Others such as the steroids may undergo chemical degradation during their circulation. Thus the oestrogenic hormone s secreted by the Graafian follicle as oestradiol. This undergoes conversion in he circulation to oestrone and oestriol, which combine in the liver with ulphates and glycuronic acid. These changes render the hormone less potent -thus protecting the organism from over-stimulation by its own oestrogen— d more soluble in water—so facilitating its excretion in the urine.

Chemical degradation

The hormone may be eliminated through any excretory channel, though it is principally sought in the urine. Even here, however, the amount present may be too small for detection, though in some cases biological or chemical methods of estimation are available. On the other hand, considerable quantities of some hormones, such as the chorionic gonadotrophin of pregnancy urine, may be excreted, the amount of hormone depending upon various

FIG 16.—The Hormones (Hormones commonly employed, in capital letters: proprietary names of commercial preparations in brackets.)

factors, such as the renal threshold, the amount in circulation and the demands of the tissues.

(3) The peripheral receptors

The thyroid and the growth hormones probably act upon every cell in the organism; others affect only certain tissues. For instance, adrenaline stimulates only those tissues supplied by the sympathetic nervous system. The tissue response may be very specific, as in the sexual skin swelling of the monkey during the oestrous phase of the ovarian cycle; here the oedematous perivulval and perianal skin is abruptly demarcated from the adjacent unaffected areas. The hormone may alter the metabolism of the peripheral cell, for example, by increasing the rate of its oxidative processes as in the case of thyroid hormone; it may facilitate the uptake of some metabolic ingredient, such as glucose in the case of insulin; or it may release some characteristic element such as calcium from the cell, as in the case of the effect of parathyroid hormone on bone.

pecific ssue esponse

3. INTEGRATION OF THE ENDOCRINE SYSTEM

The endocrine glands differ from one another in the chemical nature of their hormones, in the mechanism by which they elaborate them, and in the rate

and periodicity of hormone action, as well as in their functions. If each gland were capable of independent action the endocrine activities of the organism as a whole would be thrown into confusion. Some hormone combinations such as oestrogen and progesterone are complementary, others such as oestrogen and androgen are antagonistic; at one time a hormone may play a negligible part, at others a vital role, in the endocrine community, whereas hormones may vary in concentration and activity according to the changes in metabolism—carbohydrate, mineral or general—which they help to bring about and control. There is thus an essential need for effective integration of the endocrine system.

Compleme[ntary] and antagonisti[c] combinatio[ns]

(1) Trophic hormones

The anterior pituitary ("the leader of the endocrine orchestra") plays the most important part in this integrative process. The three glands which come especially under its control are the thyroid, the adrenal cortex and the gonads, and it regulates their activities by means of the thyrotrophic, adrenocorticotrophic and gonadotrophic hormones.

Importance of anterior pituitary

(2) Reciprocal endocrine activity

The pituitary, however, is not permitted unrestricted control of these or other glands, lest by constantly goading them it should exhaust their resources. The secretion of their hormones leads in turn to inhibition of the corresponding pituitary trophic secretion. Thus thyrotrophic hormone accelerates thyroid activity and the rate of secretion of thyroid hormone; increased concentration of thyroid hormone diminishes thyrotrophic activity. In the ovary this reciprocal mechanism is responsible for establishing a cycle of activity. Gonadotrophic hormone stimulates the follicle to produce oestrogen, which restricts further gonadotrophic stimulation of the follicle, but releases luteinizing gonadotrophin. The subsequent secretion of progesterone inhibits further luteinizing impulses and re-awakens follicle-stimulating gonadotrophic activities.

Control of effects of pituitary

(3) Hormone balance

In addition to this mechanism of reciprocal endocrine activity, examples may be given of a delicate balance set up between the complementary or antagonistic actions of two or more glands. A dog may be rendered diabetic by removal of the pancreas, but subsequent hypophysectomy will counteract the diabetes, the normal blood-sugar level being maintained by the balanced action of insulin and pituitary "diabetogenic" hormones. Hirsutism in the female is due to the androgenic effect of certain adrenal cortical hormones. These adrenal androgens are constantly being secreted, but their effect is masked in the normal woman by the secretion of oestrogens by her ovary. Mild ovarian deficiency or adrenal cortical excess may therefore give rise to hirsutism.

Hirsutism

(4) Anti-hormones

Repeated injections of hormone extracts may lead to a condition of refractoriness. This occurs only when the extract is prepared from an animal of different species from the one into which the extract is injected, and does not take place when substances such as insulin, adrenaline and the steroid hormones are used. The phenomenon is due to production of an anti-hormone

Refractory condition

in the blood of the injected animal. The anti-hormones so far identified are anti-thyrotrophic, anti-gonadotrophic and anti-lactogenic, all of them produced by pituitary extracts from species other than those in which the anti-hormone was developed. Whether these are true antibodies, or whether they are antagonistic hormones is undecided. The gonadotrophin most frequently employed for its "follicle-stimulating" properties is extracted from the serum of pregnant mares and therefore induces refractoriness when injected into the human subject, whereas the "luteinizing" chorionic gonadotrophin is derived from the urine of pregnant women and can therefore be employed indefinitely without loss of effect.

Exact nature undecided

Resistance is also developed to the continuous administration of parathyroid hormone, a substance of protein nature extracted from bovine parathyroid glands.

4. NEURO-ENDOCRINE RELATIONS

Not only should the endocrine system be integrated within itself, but its association with other systems of the organism must be delicately adjusted. Its principal relationships are with the nervous system, though hormones also take part in the control of the digestive system, for example, secretin, in the cardiovascular system, and renin in the renal system.

The most intimate connexion between the endocrine and the nervous systems concerns the peripheral effects of the autonomic nervous system. The functions of adrenaline are almost identical with those of the sympathetic nervous system, the activity of which it augments.

Role of hypothalamus

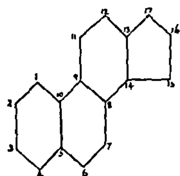
There is also a close relationship between the hypothalamus and the endocrine system. Nervous pathways may be traced via the supra-optic and other nuclei between the hypothalamus and the posterior and anterior lobes of the pituitary, and in this region certain neuro-endocrine reflexes operate to control water metabolism, the stabilization of body-weight and other metabolic activities.

Inhibitory impulses

The anterior lobe of the pituitary also responds to impulses from the higher centres, which are often inhibitory in nature. For instance, emotional disturbances and changes of environment or mode of life may lead to inhibition of gonadotrophic influences and result in amenorrhoea or alteration in the rhythm of the menstrual cycle

5. THE NATURE OF HORMONES

Chemically the hormones vary considerably in their constitution. Some are comparatively simple and of low molecular weight whereas others are of a complex nature and have not yet been chemically analysed. There are three main groups.



The cyclopentenophenanthrene ring

Cyclopentenophenanthrene ring compounds

(1) The steroid hormones

These hormones belong to the group of chemical compounds which contain the cyclopentenophenanthrene ring shown here. They comprise (a) the ovarian hormones, oestrogens

and progesterone (the corpus luteum hormone); (b) the androgens (male hormones); and (c) certain of the hormones secreted by the adrenal cortex, such as desoxycorticosterone. These hormones are soluble in oil, alcohol or ether, but not in water, and are prepared for injection in an oily medium. Table I (Preparations of Steroid Hormones) gives details of these hormones as they are at present available in the British Empire for therapeutic purposes.

TABLE I

PREPARATIONS OF STEROID HORMONES

CHEMICAL NAME	COMMERCIAL NAMES	USUAL DOSAGE	METHOD OF ADMINISTRATION
1. *Oestrogens*			
Oestrone	Unden (Bayer) Oestroform (B.D.H.) Progynon (British Schering) Menformon (Organon) Theelin (Parke, Davis)	5,000, 10,000, 50,000 i u.	Oral
Oestradiol	Unden (Bayer) Oestroform (B D.H.) Progynon B Oleosum (British Schering) Di-Menformon (Organon)	10,000, 50,000 i.b.u.	Injection
Stilboestrol		0 1, 0 5, 1 and 5 mg.	Oral
Hexoestrol		0 5 and 1 mg	Oral
Dienoestrol		1 and 5 mg.	Oral
2. *Progesterone*			
Ethisterone		5 and 10 mg	Oral
Progesterone	Lutren (Bayer) Luteostab (Boots) Progestin (B D.H) Proluton (British Schering) Lutocyclin (Ciba) Progestin (Organon) Gestone (Paines & Byrne) Lipo-Lutin (Parke, Davis)	1 5 and 10 mg	Injection
3 *Androgens*			
Testosterone propionate	Erugon-S (Bayer) Testoviron (British Schering) Perandren (Ciba) Neo-Hombreol (Organon) Virormone (Paines & Byrne)	10 and 25 mg	Injection
Methyl testosterone	Erugon-S (Bayer) Perandren (Ciba) Neo-Hombreol-M (Organon) Virormone-Oral (Paines & Byrne)	5 mg	Oral
4 Desoxycorticosterone acetate		5 and 10 mg	Injection

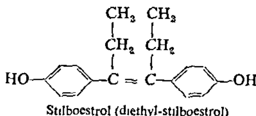
When the commercial name is the same as the chemical name the product is not listed here. Many British firms have, however, adopted the admirable procedure of describing the compound by its chemical name with the name of the firm in brackets, e g Testosterone Propionate (Boots).

Synthetic oestrogens

Though the natural oestrogenic compounds are steroids, the work of Dodds and his collaborators (1938) has shown that powerful oestrogenic activity

Substances of the stilbene group

can be achieved by administration of substances belonging to the stilbe[ne] group. This has a different and indeed simpler chemical composition tha[n] that of the natural oestrogens, *namely:*



Stilboestrol (diethyl-stilboestrol)

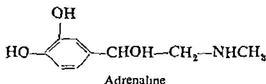
These compounds are especially active by mouth, stilboestrol being the mos[t] potent oestrogen at present available for clinical use. Two other members o[f] the group are also in common use, dienoestrol which is one-third to one-fift[h] less potent than stilboestrol and hexoestrol which is only about one-fifteenth as potent, having roughly the same degree of potency as oestrone. Stilboestro[l] is therefore the ideal oestrogen for clinical use, as far as potency, oral efficacy and cheapness are concerned. It is, however, toxic in high doses, and gives rise to nausea and vomiting in certain circumstances. For instance, women at the menopause are especially sensitive to oestrogens and complain of toxic symptoms even when comparatively small doses are given. Men and pregnant and puerperal women, on the other hand, can tolerate high doses without nausea.

Sensitivity at the menopause

(2) Simple protein hormones

Adrenaline

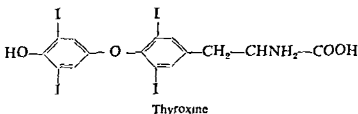
Hormones such as adrenaline and thyroxine are simple amino-acid derivatives. Adrenaline, for instance, is a derivative of the amino acid, tyrosine, and has the formula:



Adrenaline

Thyroxine

Thyroxine is also a derivative of tyrosine, and is manufactured in the thyroid cell by the iodination of tyrosine to form diiodotyrosine, with subsequent condensation of two molecules of diiodotyrosine to form a compound containing 4 iodine atoms:



Thyroxine

Though this compound is not the true thyroid hormone it has potent thyrogenic properties and is an important component of the hormone. Insulin is a more complex protein compound and has a higher molecular weight, which indicates that it is a true protein rather than a simple amino-acid derivative. It is therefore inactive when given by mouth, being destroyed by the

proteolytic enzymes of the digestive tract. This applies also to the parathyroid hormone which likewise is a protein of high molecular weight.

(3) Complex protein hormones

The anterior pituitary hormones show an even greater degree of complexity. Many of them are probably conjugated proteins, such as the gonadotrophins, which seem likely to prove to be gluco-proteins, the carbohydrate moiety playing an important part in their endocrine function. These substances have so far defied chemical analysis, though recent work with the ultracentrifuge may yield promising results. Their high molecular weight and complex protein constitution make it certain that they can never be effective when administered orally, a fact too frequently overlooked by those who advocate the use of whole-gland pituitary preparations and compound hormone concoctions dispensed in the form of expensive pills to be given by mouth.

Conjugated proteins

6. ENDOCRINE THERAPY

(1) Limitations

Endocrine therapy seldom cures. Indeed the only examples of conditions cured by hormones are certain cases of undescended testicle treated by chorionic gonadotrophin, and rare cases of menstrual disturbance in which treatment with ovarian hormones or gonadotrophins restores the normal cycle and ovulation. The principal aims of endocrine therapy are to stimulate a gland, to stand proxy for it, or to inhibit its excessive activity.

Cure rare

(a) *Stimulant treatment*

The anterior pituitary stimulates the activity of the thyroid, the adrenal cortex and the gonads. Thyrotrophic and corticotrophic hormones are seldom used therapeutically: gonadotrophins, however, have been extensively employed for many years. Gonadotrophins used for therapeutic purposes may be extracted from three sources, the anterior lobe of the pituitary gland ("pituitary gonadotrophins"), the blood serum of pregnant mares ("serum gonadotrophins") and the urine of pregnant women ("chorionic gonadotrophins"). They may be obtained in high concentration from the last two sources, and these extracts are commonly used clinically. Serum gonadotrophin stimulates the parenchymatous tissue of the gonad (the germinal epithelium) and therefore controls spermatogenesis in the male and the ripening of the Graafian follicle in the female. In these respects it behaves like the so-called follicle-stimulating hormone (F.S.H.) of the anterior pituitary. Chorionic gonadotrophin is elaborated by the placenta during pregnancy and stimulates the interstitial tissue of the gonads (indeed it is referred to in American literature as "interstitial-cell-stimulating hormone"—I.C.S.H.) and therefore controls the Leydig cells of the testis, which are responsible for producing testosterone, as well as the connective tissue structures of the cord, and the theca lutein cells and interstitial cells of the ovary

Sources of gonadotrophins

Stimulatory effects

Its actions differ, however, from those of the so-called luteinizing hormone (L.H.) of the anterior pituitary in that the pituitary hormone is intimately related to a synergic factor. In the absence of the pituitary synergist, chorionic gonadotrophin is considerably less effective. For this reason preparations are now available consisting of combined pituitary and chorionic

gonadotrophins. (For commercial preparations of gonadotrophic hormones *see* Table II.)

TABLE II

PREPARATIONS OF GONADOTROPHIC HORMONES

SCIENTIFIC NAME	COMMERCIAL NAMES	USUAL DOSAGE
1 Serum gonadotrophin	Antostab (Boots) Serogan (B.D H.) Gestyl (Organon) Gonadyl (Roussel)	200, 400, 1,000 and 3,000 i u.
2 Chorionic gonadotrophin	Prolan (Bayer) Physostab (Boots) Gonan (B.D.H) Pregnyl (Organon) Gonadotraphon S (Paines & Byrne) Antuitrin S (Parke, Davis)	100, 500 and 1,500 i.u
3 Combined pituitary and chorionic gonadotrophin	Ambinon A (Organon) Synapoidin (Parke, Davis)	

These extracts are all administered by intramuscular injection

(b) Substitution or replacement treatment

In cretinism and myxoedema

This is by far the commonest way in which hormones are used clinically. The simplest example is the use of thyroid extract to replace the deficient endogenous supply in cases of cretinism or myxoedema. Other examples are the use of parathyroid hormone in tetany, insulin in diabetes, adrenal cortical hormone in Addison's disease, and the gonadal hormones in testicular and ovarian deficiency. Substitution therapy must, of course, be continued indefinitely or until the gland spontaneously recovers its functional activity.

(c) Inhibition treatment

Atrophy of Leydig cells

Many of the glands have reciprocal effects on one another, and hormones may be employed, therefore, for their inhibitory effect on other glands. One example is the administration of oestrogens in carcinoma of the prostate to produce a castration effect. The oestrogen inhibits the gonadotrophic activity of the pituitary with the result that the Leydig cells atrophy and cease to secrete androgen. Another example is the use of thyroid extract to reduce exophthalmos after thyroidectomy. Partial removal of the thyroid gland with consequent diminution of thyroid hormone output releases the pituitary from the inhibitory effects of normal or excessive thyroid hormone levels. Excess of thyrotrophin is secreted and is responsible for the aggravation of the exophthalmos. Therapeutic administration of thyroid extract, therefore, will reduce the thyrotrophin output and consequently the degree of exophthalmos.

(2) Modes of administration

(a) Oral

The following hormones are active when administered by mouth: thyroid extract, oestrogens, especially the synthetic oestrogens, ethisterone and methyl testosterone. All other hormones either undergo destruction by the

enzymes of the digestive tract, for example, the anterior pituitary hormones; fail to be absorbed through the mucosa of the small intestine, for example, the adrenal cortical hormones; or are so transient in effect that they have lost their activity by the time they reach the blood stream, for example, adrenaline.

(*b*) *Parenteral*

Hormones which are not active when given by mouth may be administered by injection. Those which are soluble in water, such as the pituitary hormones, both anterior and posterior, parathyroid hormone, insulin and adrenaline, can be injected subcutaneously, but the steroid hormones, namely the sex hormones and desoxycorticosterone acetate, are soluble only in oil and must be injected deeply into a muscle.

Choice of subcutaneous or intra-muscular route

(*c*) *Inunction*

These lipoid-soluble hormones can also be made up as an ointment with a lanolin base or in alcoholic solution for inunction, and it is claimed that they exert a more potent local effect by this means. For instance, oestrogen ointment is sometimes used to induce mammary development.

(*d*) *By absorption through mucous membranes*

A preparation of posterior pituitary hormone is put up in the form of snuff to be absorbed through the nasal mucosa in the treatment of diabetes insipidus Some of the steroid hormones, such as the oestrogens and methyl testosterone are put up in the form of small flat tablets to be placed under the tongue or between the upper lip and the gum, where they are dissolved, with subsequent absorption of the hormone through the buccal mucous membrane. It is claimed that absorption is more effective by this route than when pills are swallowed and submitted to alimentary digestion.

Snuff

Tablets

(*e*) *Suspension of steroid crystals in aqueous solution*

Minute crystals of steroid hormones in suspension in water are injected intramuscularly. It is claimed that a prolonged effect is obtained by this method owing to slow absorption from the surface of the crystals.

Prolonged effect

(*f*) *Implantation of hormone pellets*

Considerable prolongation of effect is obtained by implanting under the skin, or deeply into the muscle, a pellet consisting of tightly compressed or fused crystals of the steroid hormones. For instance, a 100-milligram pellet of oestradiol may remain effective for more than a year, whereas a pellet of testosterone or desoxycorticosterone acetate of similar size will last for about 6 months, and one of stilboestrol for about 4 months. Progesterone can also be administered in this way, but unless implanted deeply tends to extrude through the skin. Deep implantation should be avoided in the case of oestrogens because constant administration may lead to unpleasant complications such as uterine flooding, fluid-retention obesity or even pituitary inhibition; such complications will obviously call for removal of the pellet and this can be accomplished easily only if the pellet has been placed superficially.

Possible complications

7. DIAGNOSIS

(1) History

In acromegaly and Cushing's syndrome the change in appearance of the patient is remarked upon by his relatives and acquaintances; it is sometimes noticed also in myxoedema, Addison's disease, Simmonds's disease

and thyrotoxicosis. Weight is lost frequently, though not invariably, in thyrotoxicosis; in Simmonds's disease extreme emaciation may be striking ("pituitary cachexia"), though many patients lose little or no weight. *Obesity* of characteristic distribution involving the thighs, hips, abdomen, breasts and upper arms is seen in some cases of pituitary deficiency, whereas in Cushing's syndrome the face, neck, shoulder girdle and abdomen are affected. In myxoedema general increase in weight results from the lowered metabolic activity and the thickening of the skin.

Fatigue is often an early symptom of thyrotoxicosis; muscular weakness sometimes amounting to myopathy or myasthenia is also a characteristic complaint. In acromegaly loss of muscular strength always develops in the advanced stage. Asthenia appears early in Addison's disease, and may be so severe in Simmonds's disease as to confine the patient to bed. It may be accompanied by apathy, which is found also in myxoedema, often associated with somnolence. Headaches are not characteristic of endocrine disorders, but they may become severe in cases of acromegaly when the tumour is causing tension within the pituitary fossa or, having burst from it, is giving rise to increased intracranial pressure. Both in diabetes mellitus and in diabetes insipidus there is a story of thirst and polyuria. These symptoms are occasionally noticed also in Cushing's syndrome, but when they occur in acromegaly they are usually due to the diabetes mellitus which is a common complication. Anorexia, indigestion, abdominal pain and vomiting are cardinal signs of Addison's disease, and there may be loss of appetite in Simmonds's disease. Constipation is the rule in myxoedema and may be noticed in Simmonds's disease. Diarrhoea usually develops in severe cases of thyrotoxicosis. Muscular and abdominal cramps are a troublesome feature of parathyroid deficiency. Hypoglycaemic attacks manifested by weakness and lassitude, pallor and faintness, a sensation of hunger and abdominal pain, with mental confusion, slurred speech and visual disturbances, are the chief complaint in the rare cases of islet-cell tumours, though they may be experienced in milder degree in Addison's disease and Simmonds's disease. Thyrotoxic patients tend to be intolerant to heat, whereas myxoedematous patients are sensitive to cold.

The frequent incidence of menstrual disturbances indicates how closely gonadal function is associated with other endocrine activities. Amenorrhoea is absolute in Simmonds's disease: in acromegaly, Cushing's syndrome, adrenal virilism and arrhenoblastoma, the menstrual failure, which may be incomplete, denotes excessive androgenic influence, and may be accompanied by hirsutism and acne In thyrotoxicosis scanty or infrequent periods or even amenorrhoea may occur, whereas in hypothyroidism excessive loss is more common: there is frequently no change, however, in menstrual pattern even in well-established cases of thyroid disorder In granulosa-cell tumours of the ovary and the very rare tumours in the region of the hypothalamus, occurring in childhood, menstruation may be established well in advance of the normal age of menarche Precocious sex desire is a feature of interstitial tumours of the testicle, adrenal cortical tumours, pineal and hypothalamic tumours occurring in children. Diminished libido accompanies gross gonadal failure, but may also be found in acromegaly, Cushing's syndrome and Simmonds's disease. A history of spontaneous fractures is an obvious feature of hyperparathyroidism, but is also often found in Cushing's syndrome.

(2) Physical appearance

(a) Facial appearance

Many well-established cases of endocrine disorder can be diagnosed from the foot of the bed, for in no other branch of Medicine may the facial appearance of the patient be so dramatically altered by the disease. The anxious, drawn and staring features of the thyrotoxic; the puffy, dull, complacent face of the myxoedematous patient, with bags under the eyes, scanty eyebrows and the peculiar lemon tint of the skin with a superimposed mauve-pink malar flush; the vacant, open-mouthed expression of the cretin with lolling tongue, saddle nose and wide nostrils; the square jaw, separated teeth, prominent cheek bones and coarsened features of the acromegalic; the florid, full-moon-shaped face of the patient with Cushing's syndrome; and the deathly pale and finely wrinkled skin, like crackled porcelain, of the patient with Simmonds's disease, need be seen only once, never to be forgotten.

(b) The skin

The skin is flushed, warm and moist in thyrotoxicosis; dry, scaly, pale and pachydermatous in myxoedema and cretinism. In Addison's disease pigmentation is characteristic, though other causes of pigmentation, such as sunburn, vitiligo, arsenical medication and thyrotoxicosis must be considered. The slaty-blue patches of pigmentation of the buccal mucosa are pathognomonic. The skin of the acromegalic is coarse, like normal skin seen through a magnifying glass In Cushing's syndrome there is a tendency to acne and keratosis pilaris, but especially characteristic are the striae over the lower abdomen and hips and radiating from the axilla. These are purple in colour and due to petechial haemorrhages produced by the stretching of the skin. They are unlike the pale-pink fading striae resulting from simple rapid increase in weight. The skin in Simmonds's disease is white, and suggests a severe anaemia which is seldom found. It is fine and smooth though covered with myriads of delicate wrinkles.

(c) The hair

The hair of the head is fine and glossy in the pituitary dwarf, coarse in Cushing's syndrome and acromegaly, dry and brittle in myxoedema and cretinism and poor in quality in Simmonds's disease. The eyebrows are scanty and deficient in their outer third in myxoedema, and body hair is sparse, axillary hair often being negligible. In Simmonds's disease both pubic and axillary hair are absent. Hirsutism is a characteristic feature of adrenal virilism and to a less extent of Cushing's syndrome.

(d) The voice

Speech is slow and slurred in the myxoedematous patient, and thickening of the vocal cords in this condition and in acromegaly produces a coarse and muffled sound. In advanced adrenal virilism the voice is deep and masculine. The voice of the eunuchoid fails to break.

(e) Personality

The untreated or inadequately treated cretin is imbecile; the myxoedematous patient dull and complacent, though sometimes garrulous. The thyrotoxic patient is anxious, emotionally unstable and restless; in Addison's

disease the patient is languid, and in Simmonds's disease apathetic. The eunuchoid is timid, apprehensive and retiring.

(3) Clinical examination

(a) *Anthropometry*

Engelbach's tables

There are standard tables of bodily measurements which may be used to determine whether the statural development of children is within normal limits. Those devised by Engelbach (1932) give ranges for height, weight, span, upper and lower measurements and circumferential measurements of head, chest and abdomen in relation to age. From these tables it can be determined not only whether the child is of normal height or weight, but also whether the skeletal proportions are normal. The height and span are roughly equal, and so are the upper and lower measurements. The upper measurement is from the top of the head to the symphysis pubis and the lower measurement from the symphysis pubis to the ground. The span is chiefly, and the lower measurement entirely, a measure of long-bone development, the height is half composed, and the upper measurement completely composed, of a linear measure of head and vertebral column development. Long-bone measurement depends upon the age of closure of the epiphyses, and this is chiefly controlled by the sex hormones. Premature puberty leads to early closure of the epiphyses, whereas delayed puberty results in late epiphyseal union or non-union. Thus in cases of prepubertal adrenal cortical hyperplasia or tumour, or interstitial tumour of the testicle, the span and lower measurements are significantly shorter than the height and upper measurements. In some cases of ovarian failure and in eunuchoidism, on the other hand, delayed union or non-union occurs and the span and lower measurements are disproportionately long.

Premature and delayed puberty

(b) *The pulse rate*

This is chiefly of value in the diagnosis of thyroid disorders. In thyrotoxicosis, tachycardia is a cardinal symptom and the pulse rate is between 90 and 140. In hypothyroidism it is slow.

(c) *The blood-pressure*

High systolic readings are always found in Cushing's syndrome (occasionally over 200 millimetres of mercury), in adrenal cortical overactivity and sometimes in thyrotoxicosis. In adrenal medullary tumours, attacks of paroxysmal hypertension are the essential feature. Low blood-pressure occurs characteristically in Addison's disease, in which readings below 100 millimetres of mercury are not uncommon, and also in Simmonds's disease, due to the associated adrenal cortical failure.

(d) *The breasts*

Complete failure of breast development is seen in cases of ovarian deficiency and in adrenal virilism. Atrophy of the breasts may occur to a mild degree in acromegaly and Cushing's syndrome, but is marked in Simmonds's disease. Gynaecomastia occurs in certain cases of testicular failure, especially of the seminiferous tubules. It has also been noted rarely in cases of adrenal cortical tumour ("feminizing" tumours), malignant growths of the testicle and in cases of Addison's disease treated with desoxycorticosterone acetate.

(e) *The genitalia*

The testicles should be examined in all cases of abstruse endocrine disorders in the male. Bilaterally undescended testicles give rise to the classical picture of eunuchoidism. In this condition, however, the testes may be descended but are usually small and atrophic. The testes may atrophy in acromegaly, Cushing's syndrome and Simmonds's disease, and are always poorly developed in true cases of Fröhlich's syndrome. Enlargement of the testicle, with endocrine manifestations, is usually due to a tumour, though these cases are rare. A gynaecological examination may reveal genital hypoplasia in cases of ovarian failure. In adrenal virilism there may be enlargement of the clitoris.

(4) Special examinations

(a) *Radiography*

(i) *X-ray examination of the pituitary fossa.*—This is less useful than is generally supposed. Enlargement of the fossa, with erosion of the clinoid processes or of the floor of the sella, is found only in advanced cases of acromegaly. A small fossa is usually of no significance, though occasionally a very small fossa has been reported in cases of pituitary infantilism. The skiagrams should always be stereoscopic.

(ii) *X-ray examination for bone age.*—The determination of bone age by x-ray examination of the wrist, elbow, knee and hip is of value in the diagnosis of cretinism, when bone development is retarded, and in cases of gonadal failure. In pituitary infantilism there is also retardation or complete failure of epiphyseal union. In cases of prepubertal adrenal cortical tumour and interstitial tumour of the testis there is early union.

(iii) *X-ray examination of the bone for evidence of rarefaction.*—In hyperparathyroidism typical skiagrams show generalized rarefaction or even cyst formation In Cushing's syndrome localized rarefaction may lead to collapse of vertebrae, especially in the cervico-dorsal region, giving rise to kyphosis.

(b) *Basal metabolic rate.*

The estimation of the basal metabolic rate is of considerable value in the diagnosis of thyroid disorders. Low readings are constantly found in myxoedema and Simmonds's disease. In thyrotoxicosis, however, though the basal metabolic rate is significantly raised in severe cases, obvious signs of mild hyperthyroidism may occur in the presence of a basal metabolic rate which is apparently within normal limits. The normal range is within $\pm$ 10 per cent of the standard figure (or in European countries probably 0 to $-$ 20 per cent). Thus an individual with a normal basal metabolic rate of, say, -15 may give a reading of 0 or -5 if he develops mild thyrotoxicosis. In such cases the iodine test may be of considerable assistance. Should the basal metabolic rate drop significantly following a 3-week course of iodine the patient is thyrotoxic.

(c) *The electrocardiogram*

The low-voltage pattern with poor definition of the P-waves and T-waves is characteristic of the heart in myxoedema In other endocrine disorders secondary cardiac involvement may give rise to changes in the electrocardiogram, for example, the auricular fibrillation which complicates thyrotoxicosis.

(d) Hormone assays

(i) *17-Ketosteroid estimations.*—The neutral 17-ketosteroids are the excretion products of androgens secreted by the testis and adrenal cortex. They consist of steroid compounds with a ketone group attached to the 17-carbon atom of the cyclopentenophenanthrene ring (*see* p. 34). They are estimated by means of a chemical determination involving a colour reaction. The normal figures show a mean value of about 13 milligrams per 100 cubic centimetres for women and 18 milligrams per 100 cubic centimetres for men. The ranges for female and male values are from 4 to 23 milligrams and from 9 to 28 milligrams per 100 cubic centimetres respectively. Very low values are found in Simmonds's disease, and values below the normal female range are usually obtained in Addison's disease and myxoedema. In adrenal cortical tumour and in adrenal hyperplasia the values are usually over 90 milligrams, whereas in benign adrenal virilism they may be in the region of 30–45 milligrams. In arrhenoblastoma they may be raised. In acromegaly the figure is often within normal limits except during an exacerbation, when it rises. In Cushing's syndrome the neutral 17-ketosteroids are usually within the normal range, but evidence of the presence of another corticosteroid may be found.

(ii) *Gonadotrophins.*—The presence of abnormal amounts of chorionic gonadotrophin may be demonstrated by means of the Aschheim-Zondek pregnancy test. Apart from its application in pregnancy and its complications this test is of value in the diagnosis of testicular tumour. If chorion-epitheliomatous elements are present, a positive Aschheim-Zondek test will be obtained. Evidence of the presence of excess of follicle-stimulating hormone may be found in cases of seminoma.

Aschheim-Zondek test

(iii) *Oestrogens.*—The most reliable estimation of oestrogens is obtained by biological assays, and is a complicated and laborious procedure. Oestrogen assays are of value in the diagnosis of granulosa-cell tumours of the ovary.

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HYDATID DISEASE

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1. DEFINITION

187] Hydatid disease is due to the development in man, acting as an intermediate host, of the cystic larval form of a small tapeworm, *Echinococcus granulosus* (Batsch), the normal habitat of which is the ileum of the dog. Its incidence in any country depends upon the close association of man with the dog, the definitive host, which is in turn infected by ingesting the cystic larval form most commonly found in the viscera of the sheep.

2. LIFE-CYCLE

Following ingestion the small ovum, about 40μ in diameter, hatches into a motile hexacanth embryo, which makes its way by the portal vein to the liver, where it usually enters upon its intermediate or larval stage.

Hydatid follicle

The first step is the development of a hydatid follicle, the host tissues reacting with an accumulation of mononuclear and eosinophil cells; this occurs

within a few hours. From experimental observations it has been demonstrated that this reaction often overcomes the parasite and causes its disintegration and phagocytosis. If the parasite survives it grows rapidly, and at the end of a few weeks shows vacuolation with the formation of a laminated cuticle surrounding specific hydatid fluid.

Development of cyst

As the semi-permeable, laminated, hydatid cuticle develops, leakage of specific parasitic protein ceases, the cellular reaction dies down and toleration of the parasite by the host is established, the periparasitic cells being gradually converted into fibroblastic tissue. In 3 months the cyst is about 1 centimetre in diameter, while at the end of a year a cyst about 4 centimetres in diameter, surrounded by a tough avascular adventitia, is present.

The typical simple cyst shows an outer laminated layer of hyalin and a thin inner cellular or germinal layer surrounding specific hydatid fluid—the whole being enclosed in a fibrous adventitia derived from the host.



FIG 17—Hydatid cyst of the kidney, showing daughter cyst formation

The laminated membrane is laid down from within out by the nucleated germinal layer which lines its inner aspect. It is very elastic, tends to evert itself when torn, acts as a support for the cyst and, owing to its semi-permeable nature, retains the specific fluid at a relatively high tension and protects the parasite from the entry of noxious substances.

The specific hydatid fluid is also elaborated by the germinal membrane; it is usually crystal clear, contains little protein but up to 0 2 per cent of sodium chloride and acts as a buffer and a nutritive medium for the developing scolices.

Scolices

The scolices are a sign of completed biological development; when swallowed by the dog they become active and rapidly develop into adult worms in the small intestine, thus completing the life-cycle. Unlike those of the other cestodes, they are developed within small brood capsules attached to the germinal membrane, and may be present in thousands in a fertile cyst. They are just visible to the naked eye, measuring up to 160μ in the resting state, and may be found in all stages of development from an undifferentiated cellular bud to a mature scolex with suckers, hooklets and contractile tissue.

Brood capsules

Daughter cysts

Frequently in man, who is a long-lived host, the formation of daughter cysts occurs (Fig. 17). These are small replicas of the original mother cyst and

their development is due to some interference, often by trauma, with the integrity of the original laminated membrane, so that it should be regarded as a protective phenomenon—the germinal elements being stimulated by adverse conditions to elaborate a secondary protective cuticle in order that the development of the delicate reproductive elements may go on and the species be preserved (Dévé, 1918; Dew, 1926).

3. DISTRIBUTION OF CYSTS IN THE BODY

Carriage by blood stream

The majority of hydatid cysts in man are found in the liver, this organ receiving the embryos from the portal area first and acting, as it were, as the primary filter. Sometimes the parasites pass through to the lungs where another 10 per cent lodge, the remainder making their way to other parts of the body. They may occur in any region of the body, and the lessening proportional incidence as the periphery of the body is reached indicates that distribution by the blood stream is the only rational explanation. The following Table gives the commonly accepted distribution of primary cysts.

DISTRIBUTION OF PRIMARY HYDATID CYSTS IN MAN

	ADULTS	CHILDREN
Liver	76·6	76·2
Lung	9·4	11·1
Muscular and subcutaneous	5·2	3·2
Kidney	2·3	1·2
Spleen	2·1	1·1
Bones	0·9	0·1
Orbit	0·7	1·0
Brain	0·6	4·3
Other sites	2·2	1·8

Probable frequency of pulmonary cysts

The distribution in children with its higher incidence of pulmonary and cerebral cysts probably gives a more accurate idea of the true distribution of primary cysts. The figures are taken from hospital statistics and, in view of the fact that a number of pulmonary cysts are expectorated with natural cure without the patient ever receiving medical attention, it is certain that the true incidence of pulmonary cysts is higher than that given—probably nearer 16 per cent.

Multiple infestations

It is important to emphasize that multiple infestations are much more common than is usually recognized—more than one cyst being present in at least 60 per cent of cases.

4. THE UNCOMPLICATED CYST

(1) General considerations

Age of these cysts

Simple, uncomplicated cysts are found typically in children and young adults, and their outstanding feature is their latency. Much clinical and pathological evidence shows that most hydatid cysts have been present since early childhood—the period of easy infestation—and so are nearly as old as the patient.

Toleration of cysts

Such cysts have been recorded in practically every organ, and although their slow growth allows of remarkable compensatory effects, so that huge cysts may be well tolerated, sooner or later they cause some pressure effects.

Varied and sometimes bizarre symptoms can be produced, and even small cysts in special situations, such as the spinal canal or orbit, may be serious.

(2) Uncomplicated hepatic cysts

(a) Clinical picture

The right lobe is involved in 80 per cent of all cases, and the cyst projects from the abdominal aspect in 70 per cent of these cases. Very large cysts can exist in the upper quadrants where the protected site enables them to grow quietly for many years without causing obvious symptoms (Fig. 18).

Latency

Pain is rare, and when it occurs it is nearly always indicative of the onset of some complication. Gastric symptoms are sometimes exhibited but are, as a rule, vague and not serious. Dyspnoea, which may be extreme in the case of large subdiaphragmatic cysts, is not uncommon.

Dyspnoea

Examination may reveal a rounded, tense, non-tender cystic swelling, continuous with the liver dullness, which moves with respiration. Cysts of the left lobe tend to descend anterior to the stomach, whereas those of the right lobe tend to be placed more posteriorly. In other cases the only finding may be hepatomegaly with varying degrees of increase of hepatic dullness upwards. In these cases, which tend to occur in older subjects, the amount of diaphragmatic distortion and elevation may be great, and simulation of an intrathoracic lesion may be close.



FIG 18 —Simple cyst of liver in a position notorious for its latency, also showing brood capsules attached to the inner wall.

Radiography is of great value, as it may show changes in the diaphragmatic shadow or partial calcification of the adventitia.

Simulation of intrathoracic lesion

(b) Differential diagnosis

In young subjects the findings are usually so typical that no difficulty arises, but in older subjects hepatomegaly from other causes, cirrhosis of the liver and cystic swellings of the upper abdomen, such as hydronephrosis, pancreatic cyst and mucocele of the gall-bladder, may cause errors. Radiography is essential for the differentiation of cysts of the upper quadrants from intrathoracic lesions, and, except where there is a coexistent pleural effusion, it is

Value of radiography

very accurate. In addition, the immunological reactions, particularl[
Casoni test intradermal Casoni test, give a very high percentage of positive findin
hydatid disease.

(c) Operative treatment of hepatic cysts

In those cysts which arise from the lower surface of the liver the inc
Site of incision should be made directly over the cyst, so as to give as direct an approa
possible. In those cysts which are hidden under the ribs and are high up u
the diaphragm, much care should be taken to avoid contamination o
pleura. As a rule, these large cysts can be attacked either in front or
behind. The incision should be kept as low as possible and two ribs rese
the eighth and ninth anteriorly or the ninth and tenth posteriorly. After t
Care of pleura done the free edge of the pleura can practically always be stripped u
gauze, and so a fairly large area of diaphragm is exposed through whic
incision can be safely made down to the liver.

The cyst with its white adventitia usually presents on the surface and ca
readily exposed for evacuation. It is essential that contamination of the
Risk of scolex implants of operation by active scolices be prevented, because they can implant
develop later into secondary cysts. This can be done by accurate pac
and by the partial aspiration of the cyst, followed immediately by replacer
Formalin with pure formalin. The amount of formalin should rightly be calculate
make a 1 per cent solution within the cyst, but for practical purposes
large cysts do not require more than 10 cubic centimetres of pure form
If this is allowed to act for a few minutes it kills the scolices. Although for
lin cannot penetrate daughter cysts, it still should be used in multivesic
cysts, in order to safeguard against scolices spilt from ruptured daug
cysts. The cyst can then be safely opened and the contents freely evacu
by means of a large-bore pump, and the cavity dried out. The value o
efficient suction apparatus in those cases cannot be over-estimated,
throughout the operation it should be in constant use to prevent contam
tion of the field.

Treatment of cavity The treatment of the cavity varies; if the surgeon is satisfied that the cy
clean and sterile it should be filled with warm normal saline solution to d
out the air, and closed by suture without drainage. It is an advantage to le
one long suture leading out through the incision as a guide in case of infec
manifesting itself during the next few days, when secondary drainage i
Use of drainage have to be instituted. In any suspicious case a large-bore rubber drain sho
be inserted in a suitable position; in some cases a second dependent oper
may be necessary. In large cysts, because of the risk of irregular collapse of
cavity with loculation, care must be taken not to remove the drainage t
too soon (Dew, 1929; Barnett, 1938).

(3) Uncomplicated pulmonary cysts

(a) Clinical picture

These, owing to the loose vascular tissue of the lungs, generally have a t
Situation adventitia and grow relatively quickly, so that they are usually found
young subjects. The typical cyst is univesicular, containing scolices but
daughter cysts, and may occupy either a deep parabronchial or a periphe
situation—sometimes almost filling the chest cavity. The base of the ri

lung is involved more than the left in the proportion of 60 to 40 More than one cyst is often present, and pleural adhesions are not usual.

Latency They are remarkably latent, and not more than 25 per cent are discovered before the onset of some complication. It is surprising how large a cyst can thus be tolerated. Sometimes the first indication of their presence is delay in the resolution of some respiratory infection like bronchitis or pneumonia, while a number are discovered unexpectedly during routine examination. *Cough* A dry cough is common, seems to be independent of the site of the cyst, and is accompanied by varying degrees of haemoptysis at some stage or other in *Haemoptysis* about 60 per cent of cases. There is no pain and little, if any, dyspnoea even with very large cysts. On examination, palpation rarely reveals any cardiac displacement—an important point in the differentiation from pleural effusion.

Danger of puncture A warning must be given against the use of diagnostic puncture in hydatid cyst of the lung. Owing to the brittle nature of the laminated membrane, the high intracystic tension, and the fact that a bronchial communication is only prevented by a thin layer of tissue, sudden release of pressure may precipitate intrabronchial flooding, with fatal results. Vocal fremitus is diminished over any area which impinges on the chest wall, while other signs suggestive of an intrathoracic tumour may be detected.



FIG 19 —Skiagram showing simple hydatid cyst of lung in a patient aged 43 years

(*b*) *Differential diagnosis of pulmonary cysts*

Phthisis is the arch simulator—but the basal situation of the lesion and the lack of confirmatory signs of tuberculosis should lead to its exclusion.

Great value of radiography Radiography, owing to the fact that the saline content of the cyst is relatively radio-opaque, has revolutionized the diagnosis of these cases, and great accuracy as regards both the size and the localization of the cyst is now possible (Fig. 19), although the presence of a pleural effusion may at times introduce diagnostic difficulties.

(*c*) *Treatment of uncomplicated pulmonary cysts*

Owing to the fact that a large percentage of deep parabronchial cysts undergo natural cure by expectoration, and because they are difficult to operate upon without risk, they should be treated expectantly.

Operation is, however, indicated in all cysts affecting the peripheral parts of the lung. As a rule, a cyst of any appreciable size will impinge on the parietal pleura. *Localization* It is important to localize the cyst accurately by radiography with the patient in the same position as he will assume on the operating table. This cannot be done too carefully, because upon it depends whether the operation and the post-operative course will be free from difficulty and worry. *Adherent cases* Any type of modern anaesthesia can be used, but inhalation anaesthesia by one of the

gaseous anaesthetics is best. After rib resection the pleura is usually found to be non-adherent, although in some fortunate cases it is obliterated over a rounded area corresponding to the bulging cyst. In the latter cases the cyst can be opened and evacuated boldly. There is no need to aspirate the cyst; in fact, there is danger of bronchial flooding if any form of exploratory puncture through a narrow-bore needle is used. In those cases in which there are no pleural adhesions it is advantageous to use a form of combined large trocar and suction bell, such as introduced by Fitzpatrick (1945), which allows pump evacuation to be carried out without the risk of pleural contamination—the real risk of the operation—while the lung can also be drawn well up to the surface by the suction bell. Because of the usual close proximity of a bronchus, slight intrabronchial leakage often occurs, and this is the main reason why it is inadvisable to use formalin in these cases. Full protection of the pleura and operative field by accurate packing, the use of a pump and rapid evacuation of the cyst precludes much risk of contamination of the field by scolices. After evacuation of the parasite, haemostasis is secured, a de Pezzer type of catheter sutured into the cavity, the chest closed and some form of negative pressure drainage instituted. If there has been much pulmonary collapse during the operation, it is advisable to reduce the pneumothorax, and if there has been any soiling of the pleural cavity, the use of a small intrapleural drainage tube with slight negative pressure is often advisable. Mild infective pleuritis, valvular pneumothorax and surgical emphysema are possible post-operative complications, but they are rarely dangerous and are handled in the ordinary manner.

Use of suction bell

Formalin not used

Treatment of cavity

Post-operative complications

(4) Other uncomplicated cysts

As these may occur in any organ it is impossible to describe them all. The symptoms depend not upon the nature of the parasite but upon the situation of the cyst, which acts like any other slowly growing tumour. In the same way the treatment of them follows the same principles, although in the case of special organs like the brain special techniques are indicated (Dew, 1934).

5. THE COMPLICATED CYST

(1) *General pathological consideration*

Ultimately, no matter what the situation, complications occur, their frequency increasing as age advances and as the cyst increases in size, so that in the adult the majority of cases seen in hospital are complicated. They are relatively rare in young subjects, manifesting themselves usually between the ages of 25 and 40 years. Practically all complications depend upon an escape of fluid from the cyst, this varying from a slight leak, often masked by other symptoms, to a frank rupture.

Age of onset of complications

(*a*) *Rupture of the cyst*

As the cyst enlarges, it may encroach on a natural channel, a hollow viscus or a serous cavity, one area of the laminated membrane thus becoming relatively unsupported. As a result it gives way spontaneously, even during sleep or following muscular movement, coughing, straining, or more commonly following direct trauma such as a blow, a fall, a crush or a perforating injury. Owing to its peculiar grain, the tear in the laminated membrane

Mode of rupture

Types of rupture

rapidly enlarges and allows the escape of the contained fluid and hydatid elements. Such rupture takes place commonly into the subcutaneous or muscular tissues, the bile-ducts, bronchi, alimentary canal, or urinary tract; into serous cavities, such as the peritoneum, pleura or pericardium; or very rarely into the chambers of the heart or large veins.

Each of these has its own characteristic picture, and as some of them may be combined in the individual case, a complete discussion is outside the scope of this article. The sequelae may be grouped together in the following way:

Sequelae of rupture

1. General: applicable to all types: (a) immediate; hydatid anaphylaxis; (b) delayed; secondary echinococcosis.
2. Special: applicable to cases of rupture into a natural channel: (a) immediate; mechanical effects; (b) delayed; death of the cyst with or without suppuration.

(*b*) *Hydatid anaphylaxis*

Most clinicians have noted the appearance of peculiar toxic manifestations following rupture or puncture of hydatid cysts. Although these are usually cutaneous, in the shape of erythematous or urticarial rashes, many other symptoms may occur, and there is no doubt that these are mainly anaphylactic, due to the sudden absorption of specific protein in a sensitized patient. Clinically, whenever vague symptoms arise during the course of hydatid disease, the question of whether or not they are anaphylactic in nature should always be considered (Dew, 1927).

Hydatid rash

Frequency of anaphylaxis

(*c*) *Secondary hydatid cysts*

Rupture of a hydatid cyst, or puncture by a trocar with evacuation of the specific fluid, does not necessarily cause the death of the parasite; this probably rarely occurs unless infection takes place. The parasitic elements have such powers of persistence that if aseptic conditions pertain they can survive, and ultimately develop into new cysts, often at a distance from the original; this phenomenon being called secondary echinococcosis.

Parasitic persistence

At first, clinicians and pathologists regarded all cases of multiple cysts as due to multiple primary infestations, but they gradually recognized that many were due to secondary cyst formation from implanted scolices. Even as late as 1900, however, the view that such highly differentiated structures as scolices could, as it were, revert in their life-cycle, ranked with many authorities as a biological heresy, as it seemed contrary to all the laws of the development of the cestodes laid down by van Beneden. Much experimental work and many correlated clinical and pathological investigations, however, have proved conclusively that such a retrogressive metamorphosis is not only possible, but relatively common, and of great clinical importance. Recognition of this has led to great advances in our understanding of the pathology of hydatid disease, and the following types illustrate this.

Secondary cysts

Frequency

Localized secondary echinococcosis.—A common type follows rupture of a subcutaneous or muscular cyst when congeries of small secondary cysts, often called seed hydatids, are commonly found years later. Of a similar nature are those cases of post-operative or post-traumatic recurrence in or near the site of the original cyst—one type of local daughter cyst formation.

Secondary cysts of the peritoneum.—These are very common and follow the leakage of scolices from a primary cyst of the liver or, more rarely, of

the spleen, kidney or omentum. When the cyst ruptures, hydatid fluid and scolices are shed into the peritoneal cavity with the production of some anaphylactic shock. The scolices may be shed in thousands and are carried by the rush of fluid, by gravity or by intestinal movement, as a rule to the lower quadrants. They soon become surrounded by lymph and eosinophil leucocytes and are rapidly fixed in the new site. Some are overwhelmed and undergo fibrosis; *sometimes the peritoneal reaction is so great that* a pseudo-tuberculous appearance is produced. Many, however, survive, become surrounded by a new adventitia, undergo vesiculation, and develop into secondary cysts. The peritoneum gradually spreads over until the secondary cyst gives the appearance of having developed extraperitoneally. This simple process was not understood by many of the earlier pathologists, who believed that these cysts were multiple primary cysts developed from numbers of hexacanth embryos.

Development of secondary cysts

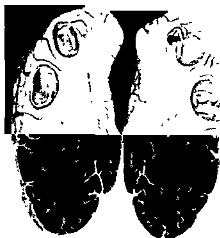


FIG 20—Multiple metastatic hydatid cysts of the cerebral hemispheres secondary to left-sided intracardiac rupture.

Pleural and pericardial cysts

Similar secondary cysts may develop in the pleural or pericardial cavities, following the appropriate rupture of a pulmonary or cardiac cyst.

Venous metastasis

Metastatic secondary echinococcosis.—This is due to a rupture of a fertile simple cyst into the heart or venous system. The rupture may take place on the venous side of the circulation into peripheral veins or into the right cardia chambers, or into the arterial sid when it takes place into one of the lef cardiac chambers. Two sets of sequela are possible. In both, of course, grav anaphylactic symptoms occur, although as a rule, recovery from the primar rupture takes place. If the cyst ruptures into the venous side the scolices an carried through the right ventricle into the lungs, where they are filtered out and give rise to secondary pulmonary cysts. These cysts are characterized by their multiplicity, their bilateral peripheral distribution, and by their uniform size.

Arterial metastasis

In the case of rupture into the left auricle or ventricle, the scolices enter the systemic circulation and give rise to metastatic cysts in various parts of the body. Owing to the position and relative size of the carotid arteries, most of the scolices are carried to the brain, which becomes the seat of the majority (60–70 per cent) of the secondary cysts (Fig. 20). The remainder of the scolices give rise to secondary cysts in the kidney, spleen or liver.

These examples illustrate the classical manifestations of secondary echinococcosis, *which is now clearly established as regards* its aetiology, pathology and clinical aspects. It is a general rule in the case of multiple cysts that if the extrahepatic cysts are more than one-third of the total it is probable that they are secondary cysts.

(d) Mechanical effects of rupture

These are seen when cysts rupture into such channels as the bile-ducts, bronchi or ureter. The entry of fluid may pass unnoticed although some anaphylactic symptoms can occur, but the debris may produce colic and partial, intermittent or complete obstruction.

(e) Suppuration

The normal laminated membrane resists the passage of bacteria, so that some degree of rupture is necessary before infection can reach the interior. Rupture causes collapse of the membrane, lowered intracystic pressure, and the entry of serum, the whole making an excellent medium for bacterial growth.

Infection of cyst

Infection is then an ever-present risk of an opening into a duct or hollow viscus, although it may be delayed for a long time, and even when it occurs it varies greatly in its effects In the case of an opening into a small duct asepsis may be maintained and the opening may heal, enabling reactive daughter cyst production to occur. When suppuration occurs the clinical picture changes to that resembling an abscess of the region.

(2) Complications in hepatic cysts

(a) Rupture of a hepatic cyst into the biliary passages

Frequency of intrabiliary rupture

As a hepatic cyst continues to enlarge, it often causes a quiet pressure necrosis of the wall of a large bile-duct, and if the opening becomes large enough some of the contents are forced into the duct. Such a rupture is relatively frequent and is, I believe, the commonest complication of hepatic cysts. It, of course, varies from a quiet leak with the passage of fluid and small particles of debris to a massive rupture, very often with occlusion of the main ducts by daughter cysts, producing varying grades of jaundice. This rupture usually takes place into the intrahepatic ducts, the passage of the slippery, non-irritating debris into the duodenum being helped by the intracystic pressure, abdominal and diaphragmatic movement and the increased flow of bile after meals (Fig. 21).

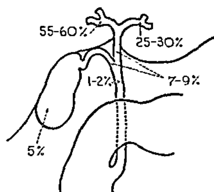


FIG. 21.—Diagram showing sites of intrabiliary rupture of hepatic cysts

Jaundice

Simulation of gall-stones

Fractional evacuation of even large cysts through the bile passages may occur over a period of many years, and the clinical picture may easily be mistaken for gall-stones in the common duct. As a rule the lower age of the patient, the predilection of hydatid disease for the male sex, the rarity of a previous history, the presence of hepatomegaly and the intermittent non-progressive nature of the jaundice and pain make one suspect hydatid disease. This may often be confirmed by finding hydatid debris in the faeces, by radiography and by the immunological reactions.

Examination of faeces

Infection

The most important risk of this particular complication is the introduction of infection. This causes suppuration in the cyst cavity, and the clinical picture changes to one of hepatic abscess If, as is often the case, the organisms are

anaerobic or virulent streptococci, these patients develop a grave toxaemia suggestive in some cases of an infective hepatitis. Others which have a safety-valve-like opening into the bile-duct may show much less toxaemia, and resemble rather a low-grade cholangitis.

Drainage of cyst

Treatment of intrabiliary rupture.—This depends largely upon the state of affairs found and upon the judgement of the surgeon at the time. Often one can attack them during a non-icteric period, when simple drainage of the offending cyst by a large drainage tube is the first essential. The cyst may be partly evacuated and therefore flaccid, and it may be deeply placed and rather inaccessible. In those cases in which jaundice is persistent it is essential to open the common duct, evacuate hydatid debris, and drain.

Suppurating hydatid of the liver should be treated like a hepatic abscess, localized accurately and drained by the most effective route. Sometimes in such cases one finds that the institution of aerobic conditions following drainage allows the florid growth of previously suppressed streptococci, and chemotherapy must then be used actively.

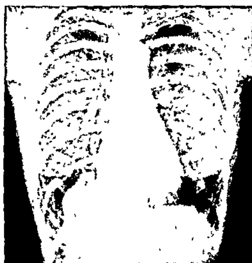


FIG. 22.—Skiagram showing subdiaphragmatic hepatic cyst extending into thoracic cavity. (Taken by Dr. H Sear a few days before intrabronchial rupture occurred.)

(*b*) *Intrathoracic extension of hepatic cysts*

Invasion of thorax

Owing to their position near the bare area and to the negative intrathoracic pressure, some hepatic cysts tend to extend into the thorax, and may ultimately rupture either into the pleura directly, or through the medium of basal adhesions and pressure necrosis of the lung into a large bronchus (Fig 22).

In its simplest form this complication produces a hepatobronchial fistula with the expectoration of bile-stained hydatid debris. A great variety of pathological and clinical pictures are [illegible]
t[illegible]
are likely (Dew, 1928).

In the treatment, the first essential is to drain the hepatic cyst, but infections of the pleura, whether ordinary pyothorax or, at the other extreme, cholepyopneumothorax, must be also dealt with.

(*c*) *Rupture into the peritoneal cavity*

Choleperitoneum

This may be produced in many ways, but injury is the most common. The effects produced vary with the state of the original cyst. If this is near or in communication with a bile-duct an immediate or delayed choleperitoneum may be produced, while in rare cases an immediate or delayed peritonitis may develop.

Secondary peritoneal cysts

Typically, however, the patient recovers from the rupture very quickly, but the scolices give rise to multiple secondary cysts, irregular in shape and size,

which owing to the tenuity of their adventitia are liable to further ruptures with repetition of the process. In this way the whole cavity may become packed with secondary or tertiary cysts of varying size (Fig. 23) This condition is often known as hydatidosis. The rent in the hepatic cyst may close, and sufficient scolices may be retained therein to produce daughter cysts in the old site later. It is therefore a general rule that whenever multiple cysts of the peritoneum are present there is also a multivesicular cyst of the liver.

Hydatidosis

Clinical picture.—The initial rupture may be quiet but is usually accompanied by severe anaphylactic symptoms, particularly urticaria and pruritus associated with some shock, pain and vomiting. The patient usually recovers rapidly and appears normal in a few days. As this may occur in young subjects the condition has often been diagnosed as food poisoning, colic, or some other transient abdominal disturbance. As a rule the whole episode is forgotten very quickly.

Anaphylaxis



Fig 23 —Multiple secondary cysts removed from omentum about 10 years after the intraperitoneal rupture of a hydatid cyst.

Variation of picture

After the lapse of from 10 to 15 years the patient begins to notice some enlargement of the abdomen, and palpation reveals the presence of multiple, rounded, tense, non-tender swellings of varying size in the abdomen or pelvis. It is obvious that the clinical picture is capable of great variation, because not only can the cysts occupy any fossa but they can in their turn undergo complications. It should be possible in practically all cases to discover the date of the original rupture years before, and often to make a retrospective diagnosis of what was at the time a puzzling condition.

Retrospective diagnosis

Treatment.—If the condition is diagnosed at the time of the original rupture exploratory laparotomy should be performed, the hepatic cyst evacuated, and as complete a toilet of the peritoneum as possible carried out. The patient should be warned of the possibility of recurrence in the form of multiple secondary cysts from 10 to 15 years later.

Treatment of initial rupture

In the established cases of multiple secondary cysts of the abdominal cavity the only treatment is patient surgical attack on each quadrant of the abdomen in turn—numerous operations over a long period of years being often needed.

Multiple operations

(3) Complications in pulmonary cysts

(a) *Intrabronchial rupture*

Intrabronchial rupture is the natural end of most pulmonary cysts, because by the time a cyst reaches a diameter of from 4 to 5 inches it nearly always comes into contact with a large bronchus, the erosion of which causes a small

part of the cyst wall to become unsupported. Following a cough, a muscular effort, an injury, or even spontaneously, the cyst wall gives way and the contents enter a bronchus. Rarely the patient is asphyxiated by the fluid or by the impaction of membrane in the glottis. More usually the dramatic symptoms are anaphylactic in nature.

Anaphylaxis

After rupture the patient usually recovers rapidly, although for some time repeated expectorations of fluid and gradual evacuation of the cyst contents may occur. He may have attacks of coughing and haemoptysis, but may remain afebrile and in good health throughout. Natural cure depends upon the size of the cyst, as upon it will depend the thickness of the adventitia, the possibility of complete collapse and the time taken for disintegration and expectoration of the cyst wall. As is to be expected, small cysts and cysts in children more readily undergo such natural cure. The larger the bronchial communication and the more dependent its position, the better are the chances of evacuation. Apical cysts are more likely to involve bronchi while yet small, and the opening will probably be dependent, facilitating drainage. Small, deeply placed parabronchial cysts also fulfil most of these conditions, and all authorities are agreed that these should not be attacked surgically and that, if left alone, up to 60 per cent will undergo natural cure. In some cases the cavity remains uninfected for long periods, while in others the presence of a fluid level with gas above—hydatid pneumocyst—gives a common characteristic radiographic appearance (Fig. 24).

Cough

Natural cure

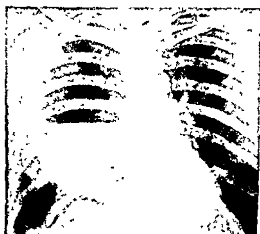


FIG 24.—Skiagram showing typical hydatid pneumocyst following intrabronchial rupture of a large pulmonary cyst; well-defined fluid level is shown.

Infection of cyst

In most cases the cavity becomes infected, at first with saprophytic, but later with pathogenic organisms, at first of low virulence, but later with virulent cocci and often anaerobic gas producers. The adventitia becomes thickened and lined on the inner aspect with a pyogenic membrane, and the clinical picture becomes one of intrapulmonary abscess, in which, owing to intermittent relief of pressure with expectoration, toxaemia varies greatly.

Treatment of intrabronchial rupture.—In the more deeply placed cysts expectant treatment is advised, but in the case of large pneumocysts or those in which infection has occurred, surgical drainage as for pulmonary abscess, by a two-stage performance in the absence of pleural adhesions, is indicated.

Two-stage drainage

(*b*) *Rupture into the pleural cavity*

The rupture of a subpleural pulmonary cyst into the pleural cavity is rare, but when it occurs it usually, because of coexistent rupture of a now unsupported bronchus, produces a special type of pneumothorax. Most cases develop a pleural effusion which contains granular eosinophil cells, while all

also run the risk of infection with the formation of a pyopneumothorax. In those cases which remain aseptic the slow development of multiple secondary cysts of the pleural cavity can also occur. This type of pneumothorax can be confused with spontaneous tuberculous pneumothorax, but, unlike that disease, it is common in adolescents, more common on the right side, there is rarely any suggestive previous history, no signs are present on the opposite side, tubercle bacilli are absent from the sputum, cutaneous anaphylactic symptoms are usual and the immunological reactions are positive.

Hydatid pneumothorax
Secondary cysts

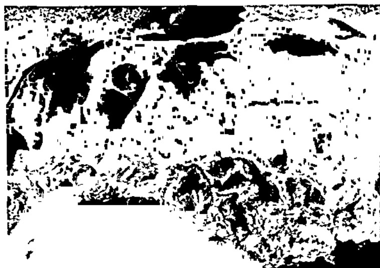


FIG 25 —Section of liver showing echinococcus alveolaris—from the first Australian case.

Treatment.—Thoracotomy with removal of hydatid debris and fluid from the cavity followed by negative pressure drainage is indicated.

(4) Complicated cysts in other situations

Space does not allow of consideration of many well-known manifestations of hydatid disease in other situations. The disease has been recorded in every region of the body, and it is obvious that the complications possible will vary in each region. Thus hydatid disease of the brain produces high intracranial pressure effects, in renal cases the cyst often ruptures into the pelvis, while vertebral hydatid disease often causes compression paraplegia.

6. HYDATID DISEASE OF BONES

In bones the growth of the parasite is greatly restricted and a peculiar diverticulated type of growth occurs. These are semi-solid buds of hyaline tissue in which little or no fluid is produced. It is only when the disease extends beyond the bones that typical hydatid cysts are formed in the soft tissues The large bones such as the femur, tibia, ilium, vertebrae and humerus are most commonly involved The growth is so slow that the disease rarely manifests itself before the age of 40 years. Spontaneous fracture, enlargement of the bone or a para-osseous abscess which simulates cold abscess

Absence of cyst formation

Clinical findings

very closely, are the outstanding findings. Radiography shows a peculiar form of rarefaction.

Operative treatment is unsatisfactory because of the difficulty of removing all the diseased tissue. Decompression laminectomy may help in the case of vertebral disease with compression paraplegia.

7. ECHINOCOCCUS ALVEOLARIS

This peculiar form of hydatid disease was first recognized in 1854 by Virchow, who discovered the parasitic nature of a disease of the liver which was formerly regarded as a colloid carcinoma (Fig. 25). For the best part of a century much controversy has raged in Europe concerning the exact aetiology and pathology of this interesting manifestation of hydatid disease. It is most common in Central Europe and for a long time was considered to have this narrow geographical distribution, but the discovery of cases elsewhere has corrected this misconception. It is characterized by its chronic infiltrating nature, its tendency to grow exogenously and break down by central necrosis, and by its ultimate fatal ending. Reference to articles by Dévé (1934) and Dew (1931) should be made for further details.

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HYPERHIDROSIS AND ALLIED STATES

By A. DICKSON WRIGHT, M.S., F.R.C.S.
Surgeon, St. Mary's Hospital, London; Surgeon, Prince of Wales's General Hospital, Tottenham, London

1. DEFINITION

188.] The term hyperhidrosis is mainly used in respect of local and pathological excess of sweating, whether idiopathic or secondary to some other pathological condition. A constitutional tendency to excessive general sweating is not regarded as pathological, although the tendency to excessive sweating in certain illnesses such as phthisis, malaria and amoebic hepatitis is part of the disease syndrome and often persists long after the primary condition has receded; frequently it is of a regional character, for example, in the nuchal region following malaria. The reverse condition of general anhidrosis, either by itself or associated with such skin conditions as ichthyosis, is a definite disease well described in dermatological and veterinary literature.

Idiopathic and secondary hyperhidrosis

2. ANATOMY AND PHYSIOLOGY

The centre for sweating lies with the heat-regulation centre in the hypothalamus and is connected with the periphery by tracts which descend in the cord slightly anterior to the pyramidal tract. From the cord, the fibres pass out in

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4. PRIMARY HYPERHIDROSIS

Primary hyperhidrosis is found in three situations: the hands, the axilla and the feet, and every situation has its special complication.

(1) Primary hyperhidrosis of the hands

There are three clinical types of primary hyperhidrosis of the hands.

(a) Hot moist hand

The hot moist hand dries only during sleep, and throughout the day is constantly soaked in perspiration; it pours with sweat under the slightest psychological disturbance. In these cases the palmar skin becomes macerated and fissured and it flakes off if the hands are used much. Palmar and digital infections may result from the fissures. The condition, seen most often in men, makes manual work an impossibility and, in severe cases, the perspiration may actually drip from the finger-tips to the floor.

Palmar hyperhidrosis

(b) Cold clammy hand

The cold clammy hand is seen more commonly in young women of the asthenic type and is sometimes associated with rheumatoid arthritis. It is a cause of acute social embarrassment and I have known it to interfere with the carrying out of certain forms of work such as nursing, piano and violin playing, stenography and draughtsmanship.

(c) General sympathetic neurosis

Cases occur in which sweating forms part of a general sympathetic neurosis of the hands. Usually the cases are of the type known as erythrocyanosis or the congestive type of Raynaud's disease. In these cases the fingers are swollen and clammy from perspiration. A very rare condition is one in which hyperhidrosis alternates with Raynaud's vasospastic attacks and even with erythromelalgic vasodilator episodes in which the fingers are hot, dry, burning and swollen.

Treatment

Whatever the type, the treatment is the same, namely, a division of the sympathetic trunk below the third thoracic ganglion and detachment of the

Sympathectomy

the anterior spinal roots and via the white rami communicantes to the sympathetic ganglionated trunk. From the ganglia, post-ganglionic fibres run in the grey rami communicantes and are distributed segmentally with the peripheral nerves. Thus it is seen that lesions of the brain, spinal cord, sympathetic chain and somatic nerves will produce disorders of sweating in corresponding areas of skin. Complete lesions produce anhidrosis, but often there is a zone of appreciable and sometimes disagreeable hyperhidrosis surrounding the dry area. In partial lesions of the nervous system hyperhidrosis is often produced in the affected regions; this is seen in causalgia and in partial lesions of the spinal cord.

The sweat glands themselves are small reddish bodies which, as a rule, lie in the subcutaneous fat or deep in the corium. When the skin is very thick the duct pursues a corkscrew course through the skin and generally opens by an oblique valve-like orifice; in thin skin the duct is nearly straight, but frequently oblique, in its course through the corium. The glands vary in size and intricacy; in regions in which the glands are sparse (neck and back) they are simple convoluted tubules; in situations in which the glands are numerous (axillae, palms, soles and forehead) they are often branched. In the axilla they form an easily visible reddish layer under the integument. The glands are said to number 2½ millions and exude on the average 2 pints of sweat per day. They are most numerous on the palms and soles, where they occur to the number of 3,000 to the square inch, whereas on the back they number only 400 to the square inch.

The physiology of sweating is peculiar, and various stimuli produce sweating in the different areas. A generally hot environment causes sweating of the whole body, but mostly of the face and the front of the trunk. The wearing of too warm clothes causes sweating in the axillae, and on a cold day the feet tend to sweat. Anxiety causes the hands to sweat, and this is the basis of the lie-detection reaction described by Waller, but stark fear makes the forehead sweat—as experienced by the surgeon in a tight corner. Shock and haemorrhage cause the forehead and face to sweat. The eating of highly-spiced curry causes only the forehead to sweat, and the eating of overheated food makes the nose shine. In the tropics the sweating occurs mostly on the trunk, but in the temperate zones the face and axillae are more affected, and on the journey home from the tropics by sea some are aware of this transition from one type of sweating to another.

Certain drugs produce sweating, for example, pilocarpine and acetyl choline which act peripherally, and pituitrin which seems to act in the same way as shock.

Numerous centres working conjointly with other brain centres must be responsible for all these different types of sweating, because they all disappear when the sympathetic nerves have been cut, except possibly the gustatory responses which may be an overflow from the secretory nerve endings of the salivary glands, and these are not innervated through the sympathetic system.

3. CLINICAL EXAMINATION

The importance of charting zones of hyperhidrosis, for diagnostic purposes, does not require stressing, and the monumental work of Guttmann (1940 (a)

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Treatment

Whatever the type, the treatment is the same, namely, a division of the sympathetic trunk below the third thoracic ganglion and detachment of the

Sympathectomy

second and third ganglia from their rami, the patient thus avoiding the disfiguring ptosis of Horner's syndrome. The anterior route is preferred and entrance into the thorax *under* the subclavian artery makes the operation much easier and quicker (Fig. 26, 27 and 28).

The incision is made parallel to, and ½ inch above, the clavicles. After defining the sternomastoid muscle its outer half is divided. The scalenus anterior

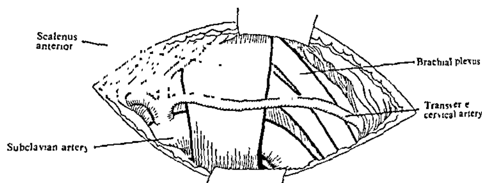


FIG. 26.—The skin platysma and outer portion of sternomastoid have been incised and the scalenus anterior displayed.

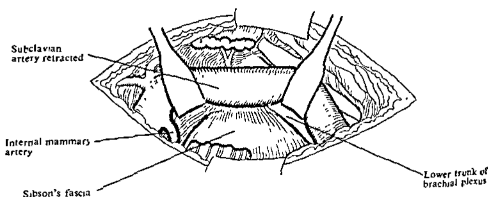


FIG. 27.—The subclavian artery is mobilized and retracted boldly upwards, exposing Sibson's fascia to its upper attachment.

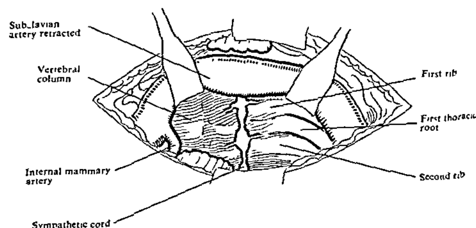


FIG. 28.—Sibson's fascia and muscle have been freely divided, freeing the whole margin of the first rib. With the lung mobilized and depressed downwards a good view of the ribs and sympathetic chain is obtained.

muscle is defined and cleaned of fat, the phrenic nerve retracted inwards and the transverse cervical artery upwards. The scalenus anter[illegible]en divided near the first rib thus exposing the [illegible]'d from Sibson's fascia and [illegible] a is di[illegible]rus. [illegible] nerve sheaths are [illegible] first rib is cleared in its entirety. A clear [illegible] of the rib is obtained and the sympathetic trunk picked up and mobilized by division of its rami. Care is taken to leave the rami of the first thoracic ganglion intact, but those to the second and third ganglia are divided, and then the trunk is cut below the third ganglion. It is best to suture the divided end to a superficial muscle to prevent recurrence. The operation may be done under local or general anaesthesia, and the use of diathermy and a sucker help to make haemostasis easier.

In all cases a most gratifying permanent result is obtained. The symptoms in Raynaud's disease recur after the operation but sweating never seems to recur in any circumstances. The operation itself causes very little disturbance and 2 or 3 days in hospital suffice. Sometimes an uncomfortable post-operative brachial neuralgia may be troublesome, but this is not seen so commonly now that the stellate ganglion is left undisturbed.

(2) Primary hyperhidrosis of the axilla

Axillary hyperhidrosis is sometimes a considerable nuisance and embarrassment, and the apocrine glands frequently over-secrete as well and add their characteristic odour. In addition, troublesome sweat rashes may affect the skin with itching and burning, and indolent infections of the apocrine glands, and axillary furunculosis may supervene.

Axillary dermatitis

Treatment

Six patients with this condition, including identical twins, have been operated on by the writer with complete success, and as all were medical students the follow-up has not been difficult. Previous to operation in some of the cases there had been considerable local treatment extending even to x-ray therapy for the troublesome axillary dermatitis. In addition the operation was performed in one case in which inveterate apocrinitis was the indication and this case also progressed very well.

The operation is the same as that for the hands except that an effort is made to get below the fourth ganglion. This is not difficult provided the scalenus anterior muscle is divided close to the first rib and the soft tissues are cleared down to the clavicle and the upper border of the first rib anteriorly.

Alternatively the approach may be made by the posterior route, after resection of the neck of the third rib.

(3) Primary hyperhidrosis of the feet

For obvious reasons excessive sweating of the feet is a more bearable complaint than either of the other two types. Epidermophytosis flourishes when the feet are moist, and macerated "soft corns" are proof of the part which sweating plays in providing suitable conditions for the life of the fungus. Although, in the main, the condition is one of the minor nuisances of life

Epidermophytosis

there are a few cases in which the disease is a major nuisance; all remedies, and there are many, fail, all activities are reduced, and holidays are spoilt. Recurring attacks of lymphangitis and cellulitis of the dorsum of the foot, and even a slowly developing elephantiasis, may make the disease a major one and the title "athlete's foot" an exasperating joke.

Treatment

In severe cases, especially if there is bromhidrosis, removal of the third and fourth lumbar ganglia on both sides should be considered. It seems rather a severe operation for such a condition, but in certain circumstances bromhidrosis is the cause of social and domestic misery. I have operated on only one case by the trans-abdominal route, and at the same time a troublesome appendix was removed. The result was perfect, and the patient's delight encourages one to consider it again, in the future, for bromhidrosis alone.

In two severe cases associated with epidermophytosis, lumbar sympathectomy was performed by the writer, and in both patients the epidermophytosis disappeared promptly. In one of the cases in which there had been recurring attacks of lymphangitis—"elephantoid fever"—these have been absent for 5 years and the elephantiasis has not progressed meanwhile.

5. SECONDARY HYPERHIDROSIS

(1) Gustatory sweating—the auriculo-temporal syndrome

In association with disease and injury of the parotid gland, there occurs after eating, a peculiar reflex sweating of the skin over the parotid gland. This would seem to be almost a perverted salivation; some substance (probably





Fig 29 Fig 30.

Gustatory sweating following successful cervical sympathectomy for Raynaud's disease. Fig. 29 shows the profuse sweating produced by the anticipation of food. Fig 30 shows complete absence of facial sweating in the hot chamber.

acetylcholine) released by the secretory nerve of the parotid gland, unable to find its normal receptor, is free to act upon the adjoining sweat glands. The condition is more than a minor annoyance, the first mouthful of food producing a rapid trickle of perspiration down the side of the face and neck (Guttmann, 1931).

The area of perspiration may, however, extend far beyond the parotid area, but is usually confined to the one side. It also happens sometimes that a small localized area on the lip or forehead sweats and the parotid region is unaffected.

Treatment

If the mechanism is that which has been described, then it is possible that sympathectomy would make the condition worse so that it would be wise to experiment with Novocain injections into the stellate ganglion before proceeding to any operative treatment, especially as one form of the syndrome follows cervical sympathectomy (*see* p 68).

Novocain injections

If the syndrome follows incision of a parotid abscess or the extensive operations now favoured in certain quarters for parotid tumours, this might constitute an argument for keeping to the older, and possibly better, operation of a careful removal of the tumour in its unruptured capsule. The conservative treatment of parotitis by massage, radiotherapy and penicillin, rather than by free incisions, would also seem to be indicated.

(2) Graves's disease

Graves's disease sometimes produces hyperhidrosis of the hands to a much greater extent than that encountered normally in this disease, even to the extent of dripping fingers. Usually the other manifestations of the disease, such as exophthalmos and tachycardia, make the diagnosis easy.

Treatment

It might occasionally happen that the thyrotoxicosis was of the "masked" variety, and a sympathectomy carried out instead of the more appropriate thyroidectomy only gives a satisfactory result as regards sweating, but leaves unchanged the other manifestations of the disease.

(3) Peripheral nerve injuries

Peripheral nerve injuries, if complete, produce a complete anhidrosis of the area of distribution, but there may be an area of hyperhidrosis surrounding the anaesthetic area. Strange to say the area of anhidrosis on one side of the body may be accompanied by an exactly corresponding area of hyperhidrosis on the other side. The value of sudorigrams in the study of nerve injuries and their reaction to treatment, needless to say, is considerable.

Partial nerve injuries

Partial nerve injuries, on the other hand, may produce a marked hyperhidrosis of the area of distribution This is well seen in a large number of cases of causalgia of the median and other nerves. The affected part may be in a perpetually clammy condition, even in a dripping state.

Treatment

In hyperhidrosis associated with partial nerve injury, sympathectomy, indicated to stop the sweating, produces in a most unexpected way cessation of the pain and an improvement in the trophic changes. The same permanent

effect is often produced by an injection of Novocain into the sympathetic ganglia concerned. If, however, only a temporary benefit is obtained resort can be had to operation. Causalgia without associated sweating does not respond so well to sympathectomy as regards relief of pain, but nevertheless it should always be tried.

(4) Trigeminal neuralgia

Cases of trigeminal neuralgia are sometimes associated with unusual sweating in the temporal area, but this does not occur so often as does lacrimation.

Treatment

The writer was encouraged in one case by reduction of pain by an injection of Novocain into the stellate ganglion; a stellectomy was then performed without improvement in either the neuralgia or the sweating. A subsequent injection of alcohol into the Gasserian ganglion cured the pain, and also abolished the sweating which had been unaffected by the sympathectomy.

(5) Sluder's neuralgia

Sluder's neuralgia is a condition which is very difficult to diagnose and to understand; it is variously described as a spheno-palatine ganglion neuralgia, a stellate ganglion neuralgia or a psychalgia. Great help can be obtained by the distribution of sweating of the face; if hyperhidrosis obtains on the affected side, or if a sweat test demonstrates asymmetry of perspiration, then the diagnosis of organic disease rather than of psychalgia can be made and operation on the stellate ganglion is indicated.

(6) Spinal injuries

In spinal injuries, disorders of sweating may be quite obvious In a complete section of the cord the anhidrosis below the injury may be associated with compensating and disagreeable hyperhidrosis above. The zone of hyperaesthesia at the trans-section level may be also a zone of constant hyperhidrosis.

The presence of a full bladder or rectum may cause profuse sweating in the upper part of the body and may be a great help to the paraplegic in the management of his urination, especially when manual expression of the bladder is the method used by the patient.

Partial division of the cord, mixed lesions of the cord and cauda equina and lesions of the cauda equina alone, produce all varieties of sweating disorders which are elucidated by the quinizarin sweat test. During the stage of recovery from spinal injury, excessive sweating may occur in the recovering areas.

(7) Following sympathectomy

After sympathectomies for vascular disorders the patient may be left with a zone round the waist in which the only functioning sweat glands are left; as a consequence all the sweating of the body is done in that region, and the constant dampness may be a source of annoyance, especially in women. It sometimes happens that, after lumbar sympathectomy, the patient complains of excessive sweating of the hands, and after cervical sympathectomy a

compensatory sweating of the feet and sometimes a gustatory type of facial sweating also occurs in areas completely anhidrotic to heat stimulus (Figs. 29 and 30).

(8) Syringobulbia and syringomyelia

Syringobulbia and syringomyelia, by involvement of the sweat tracts in the [illegible] of anhidrosis and com- [illegible] aura of sweating and [illegible] body.

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[References to other titles are given under Hyperhidrosis and Allied States in the Index Volume. The subject is also dealt with in the *British Encyclopaedia of Medical Practice* (1938), Vol. 7, p 25.]

HYPERPIESIA

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1. DEFINITION

189.] Hypertension is a clinical sign, the essential feature of which is increased diastolic blood-pressure, with which there is associated increased systolic blood-pressure.

2 AETIOLOGY

Hypertension occurs in a variety of clinical conditions, as a consequence of increased peripheral resistance to blood flow resulting from arteriolar constriction. In many of these conditions the mechanism of production of the arteriolar constriction is unknown, and in the one with which this article is concerned—essential hypertension—in the early phase of the disease no other sign or symptom may be present, and no macroscopical or microscopical lesion may be demonstrable in any tissue. Any classification of the causes of hypertension is at the present time scientifically unsound; that which follows is merely convenient for the purpose of this article.

(1) Hypertension at first not associated with demonstrable pathological lesions: essential hypertension.

(2) The specific hypertension of pregnancy.

(3) Hypertension associated with renal disease.
(a) Acute glomerulo-nephritis.
(b) Chronic glomerulo-nephritis.
(c) Chronic pyelonephritis.
(d) Polycystic kidneys.
(e) Pressure upon a renal artery.
(4) Hypertension associated with arterial disease.
(a) Arteriolo-sclerosis.
(b) Coarctation of the aorta.
(c) Periarteritis nodosa.
(5) Hypertension associated with tumours of endocrine glands.
(a) Basophil pituitary adenoma } Cushing's syndrome.
(b) Adrenal cortical tumour }
(c) Adrenal medullary tumour (phaeochromocytoma).

Some of these conditions do not lend themselves to surgical treatment; the treatment of others is dealt with in the appropriate sections. In the present article only essential hypertension will be considered.

Arteriolar constriction

The aetiology of essential hypertension is unknown. The absence of any increase in either the cardiac output or the viscosity of the blood narrows the factors to be considered to the cause or causes of the arteriolar constriction; moreover it is possible that in the earliest stages of the condition this is not present continuously, and that periods of hypertension alternate with periods in which the blood-pressure is not elevated. Vasoconstriction may result from over-activity of (sympathetic) vasoconstrictor nerves, from the action of a vasoconstrictor substance or substances circulating in the blood, or from a combination of both mechanisms.

(1) Vasoconstrictor nerves

Nervous mechanism

The wide distribution of the arteriolar vasoconstriction forces the conclusion that the only nervous levels at which such a mechanism could be mediated are the hypothalamic and cortical. While it is true that at least in the early stages of the condition the level of the blood-pressure may be reduced by the elimination of worry, by enforcing rest, by the administration of sedatives and during sleep (whether natural or induced by drugs), there is no direct evidence that over-activity of either cortex or hypothalamus is the initiating factor, and in many cases such a possibility can be eliminated. On the other hand, it will be shown that in selected cases the surgical interruption of part of the peripheral vasoconstrictor pathway may be followed by reduction in the level of the blood-pressure and by improvement in other signs, and in symptoms; however, even the most favourable operative results cannot be adduced as evidence that a direct attack has been made on the cause of the disease.

(2) Pressor substances

Humoral mechanism

Attempts to demonstrate pressor substances in the blood stream, for example by transfusing the blood of hypertensive patients to normal subjects, have been unsuccessful in man. In certain animals it has been shown by Goldblatt and his colleagues (1934) that it is possible to produce hypertension by constricting the artery of one kidney, and that this effect is not mediated

through any nervous mechanism. It is probable that the ischaemic kidney produces increased amounts of renin, small quantities of which can be obtained from the cortex of the normal kidney, and that the renin acts upon a certain fraction of plasma globulin to form hypertensin (angiotonin), the pharmacological action of which is directly upon the arteriolar walls.

Renin

Hypertensin

Though almost constant at necropsy, pathological changes in the renal arterioles of patients who have died of benign essential hypertension are rarely reflected in ordinary clinical tests; sooner or later more exact measurements of renal function demonstrate a reduction in renal blood flow due to constriction of the efferent glomerular arterioles. It is possible that this may both perpetuate and intensify the hypertensive state.

3. COURSE

According to the clinical picture they present, patients with essential hypertension may be classified into two groups.

(1) Benign hypertension

The majority of patients belong to this group; they seek advice because of symptoms—headache, giddiness, lack of concentration, lack of ability to complete sustained or sudden physical effort, and the so-called "hypertensive encephalopathies", for example temporary paralysis, temporary aphasia and temporary disturbances of vision. The cause of death is cardiovascular in over 80 per cent of cases and renal in less than 10 per cent. The possibility of surgical treatment is limited to this group.

(2) Malignant hypertension

(a) Supervening on the "benign" type, and (b) rapidly progressive from the first. Irrespective of symptoms, the striking pathological lesion is glomerular afferent arteriolar necrosis, with rapid renal failure and uraemia.

The former type is encountered in patients in later life, the latter may occur at any age. Surgical treatment is contra-indicated.

4. INDICATIONS FOR SURGICAL INTERVENTION

During the past twenty years various surgical procedures have been advocated in the treatment of essential hypertension. They have been based upon two considerations: (1) the blood-pressure is not raised in an effort to force more blood to ischaemic organs, and (2) commonly, the cause of death is attributable to the raised blood-pressure. Their objects have been three: (1) to remove vasoconstrictor tonus from as large as possible an area of the arteriolar bed by sympathectomy; (2) to reduce the production of the (normal) pressor-substance, adrenaline, by adrenalectomy or by interruption of the nerves to the adrenals, and (3) to remove vasoconstrictor tonus from the renal artery by denervation of the renal pedicle. In the light of present knowledge, no one of these objects, and no combination of them, is directed to eliminating the cause of the disease: yet operations based upon them have been attended by a proportion of at least temporary successes, not only in lowering blood-pressure, but also in ameliorating other features of the condition.

Objects

In essential hypertension, operation is to be undertaken only after the most complete physical examination to exclude any of the other causes of

hypertension included in the list on p. 71; in practice certain clinical tests may be utilized to exclude those patients unlikely to benefit.

(1) The blood-pressure

In general, operation is to be considered only when the blood-pressure is labile, and falls after rest, during sleep, or after sedation. The most useful test is to administer 3 grains of Sodium Amytal every hour for 3 doses, and to chart the blood-pressure ½-hourly between the second and fifth hours. If the diastolic pressure does not fall below 100 millimetres of mercury, a favourable result from operation is unlikely; a fall below 100 does not necessarily guarantee a favourable result, but the nearer the approach to a normal level the better the prognosis after operation.

(2) The renal function

Any primary cause of hypertension such as acute or chronic glomerulo-nephritis, or bilateral pyelonephritis, must be absent. The kidneys must be able to secrete urine of a specific gravity of 1·020 or higher, and the level of non-protein nitrogen in the blood must not exceed 40–45 milligrams per cent.

(3) Cardiovascular system

Arteriosclerosis, evidence of congestive heart failure and the occurrence of angina pectoris are all contra-indications to operation.

(4) Ophthalmoscopic examination

Papilloedema is an absolute contra-indication to operation, and the presence of haemorrhages and patches of exudate are of unfavourable prognostic significance.

(5) Cerebral vascular episodes

Encephalopathies

Patients who have experienced one or more cerebral vascular episodes may be accepted for operation only when they have made a complete neurological recovery.

(6) Choice of patients

Age

Patients should certainly be under 50 years of age and preferably under 40 years.

Other cases

If cases are selected for operation by the rigid application of these tests, it is likely that only those for whom benefit is possible will be subjected to surgical treatment. The more experienced may accept for operation certain patients who would be excluded by the tests. These are of two types; younger patients, whose response to Sodium Amytal is poor and who may have retinal haemorrhages, for whom the limited result of symptomatic relief is sought; and older patients, with the early signs of heart failure and possibly with slight renal damage, who may also be freed from symptoms by operation, and who remain relatively comfortable until they die suddenly as the result of a cerebral vascular accident. It must be emphasized that considerable experience is required in accepting such patients for operation, and that it is best for the occasional operator or the beginner in such work to be strict in his selection of cases.

There remains the question of "prophylactic" operation when transient episodes of hypertension have been discovered in a young adult: in the writer's opinion the present state of knowledge does not justify surgical treatment of such patients.

5. OPERATIVE TECHNIQUE

It is probable that the more extensive the sympathetic denervation, the more satisfactory the result. Three types of operation have been advocated.

(1) Supradiaphragmatic splanchnicectomy and sympathectomy

Supra-diaphragmatic route

The splanchnic nerves and lower part of the thoracic sympathetic chain are interrupted through an approach, devised by Peet, Woods and Braden (1940), which includes resection of the posterior portion of the eleventh rib.

(2) Subdiaphragmatic splanchnicectomy and sympathectomy

This operation is performed through a high lumbar incision, the twelfth rib being resected (Adson, Craig and Brown, 1936).

(3) Combined supradiaphragmatic and subdiaphragmatic splanchnicectomy and sympathectomy

Spinal anaesthesia to the level of the fifth or sixth thoracic segment, supplemented by nitrous oxide and oxygen, or by cyclopropane, is suitable.

The patient is placed on the operating table in the "kidney" position, with the side to be dealt with inclined 30 degrees towards the surgeon, who stands



FIG. 31—Incision for combined supradiaphragmatic and subdiaphragmatic splanchnicectomy and sympathectomy.

Incision

at the patient's back. The line of the eleventh rib is identified; the incision is made in this line, from the outer border of the sacrospinalis muscle to the outer border of the rectus abdominis muscle (Fig. 31) The superficial structures are incised down to the rib, which is then removed subperiosteally from its tip to the posterior limit of the incision, care being taken to preserve the pleura intact. The extrapleural tissues are entered at the posterior end of the incision, and separation of the pleura from the thoracic wall is begun with a finger-tip and continued with small moist swabs mounted on long artery forceps. It is important that the separation of the pleura should be carried out systematically and gradually in every possible direction—cranially, both anteriorly and posteriorly, towards the median line, and from the upper surface of the diaphragm. When a sufficient area has been separated, the pleura is retracted anteriorly and medially by a Deaver retractor, the blade of which

Separation of pleura

; covered by gauze. The field of operation is now illuminated by a headlight. ιs the dissection proceeds medially, the tenth and eleventh thoracic ganglia nd the sympathetic trunk come into view (Fig. 32). The separation of the leura follows the chain first in a cranial direction, to a level which varies with ıe configuration of the chest, and which may reach as high as the seventh ıoracic vertebra. The dissection now turns medially, at about the level of the ınth ganglion, and soon displays the roots of the greater splanchnic nerve, assing medially from the ganglionated chain. The pleura is separated towards

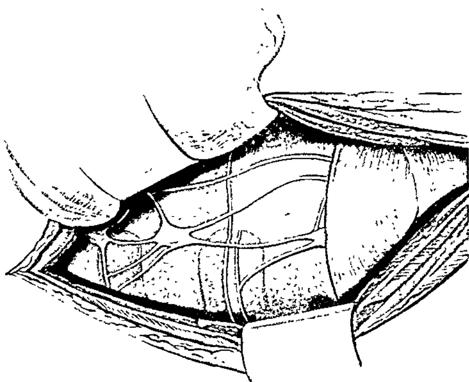


FIG. 32.—The supradiaphragmatic part of the operation. The lower part of the thoracic chain and the lower roots of the splanchnic nerves exposed by retraction of the pleura medially.

the median line from this point caudally, until the roots join the greater splanchnic nerve; the latter is then gently isolated as cranially as is possible on the one hand, and on the other to its point of disappearance into the crus of the diaphragm: for this part of the dissection long-handled hooks and long, fine, blunt scissors are convenient. The eleventh and twelfth thoracic ganglia are then displayed, the latter by separating the diaphragm by gauze dissection from its origin from the medial and lateral arcuate ligaments. Between the eleventh thoracic and the first lumbar ganglia the sympathetic chain is usually attenuated and must be dissected with care. The greater splanchnic nerve and the sympathetic chain are now avulsed as cranially as the exposure permits, and the chain is freed caudally by raising

it from the posterior thoracic wall so that the rami communicar intercostal nerves may be divided with scissors, care being take intercostal vessels are not injured. A warm moist pack is placed in th cavity, the lung is expanded, and the abdominal part of the operatio

Abdominal part of operation

The external oblique, internal oblique and transversus muscles a in the line of the incision, to expose the extraperitoneal fat, and th the diaphragm is separated from the inner surface of the twelfth extraperitoneal fat is separated from the psoas muscle as far as i

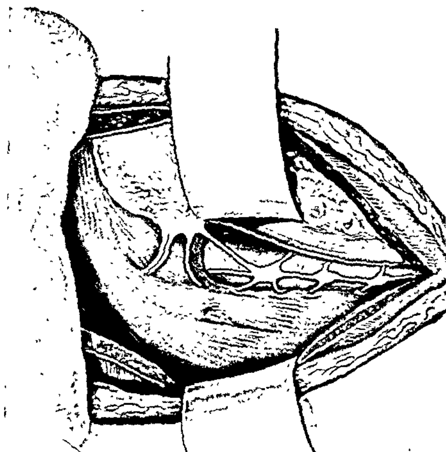


FIG. 33.—The subdiaphragmatic part of the operation. Above, the splanchnic ne joining the coeliac ganglion. Below, the lumbar chain emerges from beneath the the diaphragm. The representation is somewhat diagrammatic.

border, and the kidney in its perirenal fat and fascia is retracted media caudally to expose the abdominal aspect of the crus of the diaphragr greater splanchnic nerve is readily identified as it appears between the and intermediate pillars of the crus and passes medially and anteriorly the semilunar ganglion (Fig. 33): it is divided a short distance from the lion, and here a branch of the phrenic artery may require ligature. The splanchnic nerve is also divided (*if it can be identified*) as it pierces th phragm lateral to the greater splanchnic nerve between the intermedia lateral pillars of the crus. The lumbar sympathetic chain is then identifie avulsed below the second or third lumbar vertebra, and dissected cran the first lumbar ganglion can be isolated only after the musculo-aponer

opening in the diaphragm under which the chain disappears has been slit upwards for a varying extent.

The isolated nerve strands are now removed, either from the thoracic or from the abdominal portion of the wound. If the pleura has been punctured, a catheter is placed in the extrapleural space, and suction is maintained on it until the closure of the wound is completed. The abdominal muscles are closed in three layers, the muscles over the rib space in one layer. Attention must be given to two important points: (1) the detached origin of the diaphragm is included in the sutures uniting the posterior part of the transversus muscle, with which the diaphragm interdigitates, and the diaphragm is also sutured to the intercostal muscles of the eleventh space; (2) the eleventh intercostal nerve is not included in the suture line.

6. POST-OPERATIVE CARE

The post-operative course is usually uneventful, although there may be intercostal pain for a few days; for the first 24 hours a carbon dioxide and oxygen mixture should be breathed for 5 minutes every hour, to help to prevent atelectasis. The patient may get up about the fifth day, and a similar operation may be performed on the other side about the tenth day. After the second operation, and even although there is no post-operative fall of blood-pressure in the horizontal position, in the majority of cases there is marked postural hypotension when the patient assumes the erect position. Unless this is minimized, syncopal attacks may occur; therefore the patient should wear a firm abdominal binder, and should move about rather than stand still. This postural hypotension gradually disappears over a period of about 3 months.

Postural hypotension

7. RESULTS OF TREATMENT

When patients are carefully selected for operation, the operative mortality should be low (less than 1 per cent) The immediate results of treatment may be considered under two headings.

(1) Reduction in blood-pressure

In general it may be said that reduction in blood-pressure follows operation most often, and is of greatest range, when the pre-operative blood-pressure, and especially the diastolic pressure, was not unduly elevated before operation. In a small proportion of cases (probably not more than 25 per cent) the blood-pressure remains at this lower post-operative level for at least some years; more often the reduction is not maintained, and the blood-pressure gradually rises again over a period of months or years. In many cases there is no significant post-operative reduction of blood-pressure; in the absence of accurate knowledge of the mechanism of operative relief, it is impossible to predict the effect of surgical intervention on blood-pressure, and it is idle to speculate on the factors which lead to reduction in one case, and absence of reduction in another clinically similar case.

(2) Amelioration of symptoms

On the other hand, and even in the absence of a significant post-operative reduction of blood-pressure, a high proportion—about 75 per cent—of

patients are relieved of such symptoms as headache and giddiness. The ter ency to episodes of hypertensive encephalopathy is diminished, the cardi hypertrophy is reduced, the appearance of the retinal vessels is improved, a fatigue is less easily induced. The important result of this symptomatic rel is the ability to return to previous activity, or to activity of a somewhat l exacting form.

The influence of operation on life expectancy is as yet unknown.

8. SUMMARY

In carefully selected cases operative treatment occasionally restores t patient to a clinically normal state, and usually to a state in which, at lea temporarily, he is able to lead a happy and productive life.

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IMMERSION-FOOT

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1. DEFINITION

190] The term "immersion-foot" was coined during World War II to describe a syndrome observed in shipwreck survivors whose extremities had been exposed to the effects of cold sea-water. Immersion-foot is akin to other conditions, such as trench-foot, which are the result of prolonged exposure to low temperature insufficient to freeze the tissues.

2. AETIOLOGY

Temperature range

The essential aetiological factor is exposure of the limbs to cold. The temperature range within which immersion-foot occurs is thought to lie between sea freezing-point (−1·9° C.) and 15° C.; the lower the temperature the shorter the period of exposure required to produce irreversible changes. Immersion has no specific action other than its effect in increasing heat loss; cases of an identical syndrome have been observed in which the only moisture was the patient's perspiration; but prolonged immersion may cause maceration and thus increase tissue damage. Other important contributory factors are immobility, dependence and constriction of the limbs, malnutrition, low morale, the effect of cold upon the whole body, and haemorrhage and shock from wounds.

Immersion

Other factors

3. MORBID ANATOMY

All tissues of the limb are affected to a variable degree, but nerve and muscle suffer most severely. Immediately after exposure the skin and subcutaneous tissue show cellular and intercellular oedema, succeeded in the late stages by the deposition of fibrous tissue and collagen. In muscle the early lesion is a patchy Zenker's hyaline necrosis; later the changes are those of denervation. The nerves undergo partial or complete Wallerian degeneration followed by regeneration, damage being greatest in fibres of small calibre. Some workers have reported thrombosis in the main arteries and veins, but it is doubtful if this is a primary pathological change. The minute vessels are dilated and

Skin and subcutaneous tissue

Muscle

Nerve

Blood-vessels

tortuous; their lumina may be occluded by conglutinated red blood cells or by hyaline thrombi. Bone is the least damaged tissue but may show decalcification, followed by regeneration.

4. CLINICAL PICTURE

The clinical features of immersion-foot are best described in four phases:

(1) During exposure

The feet are cold and numb. Pain is not felt at this stage. The feet are a pale, sickly-yellow colour or blue and mottled in appearance; occasionally they may appear bright red. After a few hours swelling begins and footwear has to be removed. The cold, sodden, insensitive tissues are friable and easily damaged by minor traumas.

(2) The pre-hyperaemic stage

This occupies the first 6–24 hours of the post-rescue period. The feet remain cold, swollen and numb, and feel heavy; as a rule survivors are unable to walk without assistance; toe and ankle movements are impaired; frequently the ankle jerks cannot be elicited; there is a "stocking" type of sensory loss to all forms of exteroceptive stimuli and the peripheral pulses are impalpable.

(3) The hyperaemic stage

This stage develops with remarkable rapidity. Within a matter of a few hours the previously cold, pulseless feet become hot, and the pulses are full and bounding. Except in mild cases the onset of the hyperaemia is attended by pain which at first is of a severe, diffuse, burning or throbbing type, and increases in intensity to reach a maximum in 24–36 hours. Later shooting or stabbing pains develop; these occur in paroxysms, are aggravated by heat, by movement, and occasionally by emotional factors, and radiate from the centre of the foot towards the toes. The feet are now of a red-blue colour; swelling tends to increase; and blisters, ecchymoses and petechial haemorrhages appear. Areas that are to become gangrenous do not warm, blister extensively and are surrounded by a well-defined line of demarcation. At this stage the appearance of the feet may be most alarming (Fig. 34). Within a day or two the intense hyperaemia subsides and the feet, although still abnormally warm, become pale with areas of gangrene clearly defined. Superficial necrosis with loss of portions of the toes is not unusual, but massive gangrene with loss of more than toes is rare. Seven to ten days after rescue the area of sensory loss is of "sock" or "carpet-slipper" type, with analgesia slightly more extensive than anaesthesia. As swelling subsides, wasting and paralysis of the intrinsic muscles of the foot become apparent and contractures of the toes may follow.

Pain

Appearance of feet

Gangrene

Sensory loss

Contractures

(4) The post-hyperaemic stage

This follows in all but the mildest cases. The transition is never abrupt; the previously warm feet gradually become cold. At first coldness is transient and always the result of some cooling influence; later the feet are permanently cold and are cold-sensitive. Excessive sweating of the feet is the other troublesome symptom of this phase.

Cold-sensitivity

Hyperhidrosis

The above is a description of a typical case of immersion-foot of moderate severity. Tissue damage depends upon the temperature to which the feet are

exposed and the duration of exposure. All grades of severity from slight swelling and mild, transient hyperaemia to massive gangrene of the feet are observed.

Grades of severity

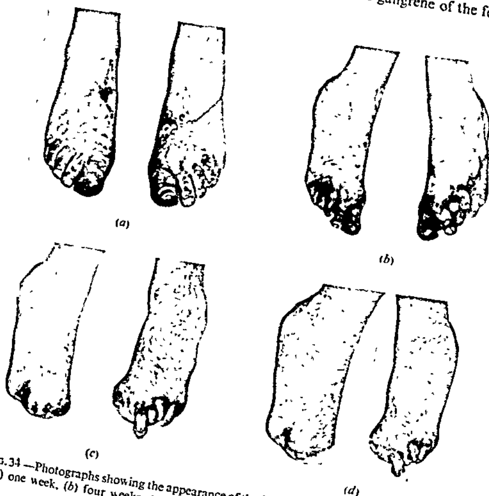


FIG. 34.—Photographs showing the appearance of the feet in a severe case of immersion-foot (*a*) one week, (*b*) four weeks, (*c*) fourteen weeks, (*d*) twenty-nine weeks after rescue

Immersion-hand.—In approximately 50 per cent of cases the hands are also affected, although usually less severely than the feet. The essential features of immersion-hand are similar to those of immersion-foot. Wasting of the intrinsic muscles of the hand gives rise to an appearance similar to the "claw hand" of progressive muscular atrophy. Attacks of the Raynaud phenomenon have been observed as a feature of the post-hyperaemic stage of immersion-hand.

5. DIFFERENTIAL DIAGNOSIS

Immersion-foot must be distinguished from true frost-bite; in the latter freezing of the tissues follows a brief exposure to severe dry cold. A syndrome similar to immersion-foot has been observed in the limbs of shipwrecked survivors adrift for long periods in the Gulf Stream; hypoproteinaemia, vitamin deficiencies and exposure to strong sunlight are the principal aetiological factors in this syndrome.

Frost-bite

Oedema in "warm-water survivors"

6. PROGNOSIS

Prognosis depends upon the extent of irreversible tissue damage as judged by the condition of the feet 7–10 days after rescue. Many patients who appear initially to have suffered only a mild degree of immersion-foot may still complain of symptoms months or years later, particularly if they return to sea in northern latitudes. Late sequelae include pain in the feet, persistent coldness and hyperhidrosis, intermittent claudication and flat-foot.

Late results

7. TREATMENT

General

After rescue survivors should not be allowed to walk; stripped of their wet clothing they should lie with their feet elevated and exposed to the air. The trunk and proximal portions of the limbs may be warmed, but the application of heat to the chilled extremities is strictly forbidden. It has not been proved that cold therapy is of value at this stage. Once the feet enter the hyperaemic stage, dry cooling *is of definite value for the relief of pain; the aim should be* to keep the temperature of the feet at 20–25° C. In the majority of cases morphine also will be required. An injection of anti-tetanus serum should always be given. The feet should be kept clean and dry; they should be washed daily with soap and water followed by spirit, dusted with sulphanilamide powder and wrapped in a sterile towel. Blisters should be snipped aseptically and necrotic tissue removed. Any question of amputation *must be deferred until* the extent of the gangrene is defined clearly. As a rule "trimming" operations will then give the patient a useful foot; formal amputations are seldom necessary. Pre-ganglionic sympathectomy may be of value for the late cold-sensitive state and hyperhidrosis, but should not be performed in the early stages. The treatment of late contractures is an orthopaedic problem.

Cold therapy

Care of the feet

Amputation

Sympathectomy

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[References to other titles are given under Immersion-Foot in the Index Volume.]

IMPOTENCE

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191.] Surgery has a very small place in the treatment of impotence because failure to complete the sexual act satisfactorily is far more often psychogenic than organic in origin. Even when a physical lesion is discovered, such as tabes dorsalis, this lesion is rarely amenable to surgical measures. Surgery is therefore mainly confined to the removal of such hindrances to coitus as gross curvature of the penis, or to the elimination of pain, such as that resulting from the existence of a tight prepuce.

Psychological effects of surgery

Although in the opinion of the writer, surgery is of little importance in the treatment of impotence, many surgical measures have been devised for increasing the potency in patients who have experienced difficulties in coitus, either because of a premature ejaculation or because of an incomplete erection. Sometimes these surgical measures are successful, but their success is often due to the strong suggestive action which they exert on the patient. Fortified by the thought that an operation has remedied the organic defect from which such patients believe themselves to suffer, their confidence in their powers is restored. This alone will often bring about a cure. A striking example of this, in my own practice, was the case of a youth who complained that he had destroyed his virility by tearing a ligament during the act of masturbation. He had previously consulted many doctors, and his condition was by then so preying on his mind that he even contemplated suicide. Realizing that it was quite impossible for him to accept any assurance that his genital organs were intact, I promised him that nothing would be easier than to unite by operation the torn ligament. Having taken the relatives into my confidence and obtained their consent, I arranged that he should be given an anaesthetic. A small incision was made in the skin over the site of the supposed injury and immediately sutured. While coming round from the anaesthetic the patient was allowed to hear a conversation in which the complete success of the operation was repeatedly emphasized. As the result of this suggestion, supplemented by further psychotherapy, he quickly regained his potency. Suggestion also played a large part in those cases of impotence which I treated by testicular grafting prior to the discovery of testosterone. Even when human grafts, obtained from cases of ectopic testes were used, they underwent rapid absorption, and the amount of hormone passing into the blood during this process was too little to have had much effect on potency.

Testicular grafting

What is true of grafting is equally true of the one-time popular Steinach operation. Yet patients prefer these "magic" operations to a tedious course of psychotherapy, for there is a stigma attached to a disturbance of the mind which is not attached to one of the body.

Fibrosis following cavernositis

As previously stated, intercourse may be rendered difficult either because it causes pain, or else because it is associated with mechanical difficulties. In curvature following fibrous cavernositis both of these hindrances may be present. This particular lesion is avowedly difficult to treat. Lowsley (1943)

has recently reported favourably on the surgical *extirpation* of the plaqu responsible for the deformity. The *operation* he recommends is as follows. *tourniquet is applied to the root of the* penis and a dorsal *incision is made* i the midline which starts at the coronal sulcus *and* passes backwards towar the pubis. The *thickened* tissues are entered *through the septum lying* betwee the two corpora cavernosa. All pathological tissue is excised, *care being* take to avoid entering cavernous tissue *more than is necessary. The* wound is the repaired, particular attention being paid to the accurate approximation c Colles's fascia, known *in America as Buck's fascia.*

Excision of plaques

The numerous surgical procedures which have at various *times been* carrie out in the posterior urethra are all based *on the supposition* that prematur ejaculation is the result of pathological changes in *this region,* and mor particularly in the region of the verumontanum. The posterior *urethra* ha indeed been the happy hunting ground *of enthusiastic* urethroscopists wh have discovered that ejaculatio praecox is associated with a swollen, congested and over-sensitive verumontanum. This is true of those cases whic follow a posterior urethral infection, but in my opinion it is not true of th generality of cases. Huhner (1937) advocates treatment by means of instillations of silver nitrate. These are made through an Ultzmann's catheter, *starting with a strength of 1 in 2,000* and working up gradually to a strength of 1 in 500. This treatment may be combined with prostatic massage, or with the passage *of a full-sized metal bougie. Other urologists recommend more* energetic measures. The verumontanum may be cauterized with silver nitrate melted on a probe, or coagulated with a weak diathermy current. I have tried all of these measures, but with indifferent success. Premature ejaculation, like *incomplete erection,* is more frequently due to psychological difficulties than to organic changes in the posterior urethra. It responds better, therefore, to psychotherapy than to surgical measures, but treatment will often have to be *prolonged.* Sometimes this form of sexual difficulty is an anxiety symptom; sometimes it is the result of a resistance on the part of the patient to commit himself to the complete act of sex.

Abnormalities of verumontanum

Psychotherapy

The commonest form of impotence is that in which the patient fails to obtain a *sufficiently rigid erection to allow of penetration,* and special operations have been devised to remedy this. The exact mechanism of erection has not yet been fully worked out. All that is known is that stimulation of the nervi erigentes is followed by vasodilatation of the arterioles of the penis, and that the turgescence thus brought about is increased by a raised resistance to the return of blood through the efferent veins. At one time ligature of these veins was practised, but with *indifferent success.* The turgidity of the congested organ is completed by compression of the efferent

of the penis

e intrinsic

an operation for enhancing the action of the ischiocavernosus and bulbocavernosus muscles. It is carried out as follows. The patient is placed in the lithotomy position and a Number 20 French metal bougie is passed along the urethra. An incision is then made in the midline over the bulging part of the perineum, starting 10 centimetres in front of the anus, and is carried backward for 5 centimetres. A branch is made laterally on each side to *points just above the*

Incomplete erection

Lowsley's operation for enhancing the action of the ischio-cavernosus and bulbo-cavernosus

attachments of the crura penis, the completed incision thus representing an inverted Y. This is deepened, and dissection is carried out until the corpus spongiosum, surrounded by the bulbocavernosus is exposed. The ischio-cavernosus muscle fibres are also identified. Chromic ribbon-gut sutures are next inserted into the lateral edge of the bulbocavernosus, pulled across the belly of the muscle, and attached to the opposite side, with just sufficient strain to plicate the muscle. The idea is to produce that amount of pressure required to reinforce any contraction, and thus to aid in producing erection. Two other similar stitches farther forward may be necessary to tighten the whole muscle. The ischiocavernosus muscles are shortened on each side by ribbon-gut sutures, and in his later operations, Lowsley aided compression of the dorsal vein by passing an additional suture through each leaf of the suspensory ligament and over the dorsal vein. Throughout the operation the ligatures must be tied without too much tension, as otherwise a persistent and painful erection may result. Lowsley claims 31 successes out of 51 patients operated upon. He states that the operation is particularly useful when the impotence is the result of previous perineal trauma with damage of the perineal muscles. He also asserts that it is likely to fail in the case of men over sixty years of age whose muscles have deteriorated and become infiltrated with fat. My own comment on these indications for operation is that all attempts to restore potency are more difficult when the patient is elderly and nearing the physiological end of his sexual life, and that impotence resulting from perineal trauma is exceedingly rare. It may well be that Lowsley's operation is successful in convincing a patient that his penis is capable of becoming erect because the tightening of these perineal muscles is often followed by a post-operative erection. In other words, suggestion probably plays a large part in Lowsley's successful results. I have little personal experience of this method of treating impotence, having performed it on only one occasion. The patient's mentality rendered him an unsuitable subject for any form of psychotherapy, and the operation failed to cure him.

Psychology has been described as a superstructure, and the physical basis on which this superstructure rests is unknown to us. It is probable that in most so-called psychogenic cases there are also biological changes, but generally these changes are unknown and, even if they were known, they would not be reversible by the crude methods of surgery. In my opinion, the surgical treatment of impotence will always remain of secondary importance.

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[References to other titles are given under Impotence, in the Index Volume. The subject is also dealt with in the *British Encyclopaedia of Medical Practice* (1938), Vol. 7, p. 103.]

INFECTION, INFECTIONS AND INFLAMMATION

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1. HOST REACTIONS IN GENERAL

192] Disease may be regarded, with profit, as a conflict between a living organism, host or recipient and an injurious agent, but something more than a conflict is implied, inseparable from the maintenance of physiological balance within the organism, and in this way the special problems of disease link up with those of biology in general. The injurious agent, though often a micro-organism from the outer world, may be closely connected with the host, in some cases being a product of the host's metabolism or arising through something going wrong in the working of the host. Hence we may consider disease in terms of mutual relations between host and agent and these relations fall into place when time sequences are adjusted. The initial association may prevent the agent from making further contact with the host, through measures exerted by the latter or perhaps because of a lack of aggressiveness on the part of the agent. Then there is the possibility that the agent, having gained entrance to the host, may be prevented from penetrating far into the tissues. A further stage is reached when the agent, having broken through

Relation between host and agent

these barriers or, as sometimes happens, having been carried past them by some mechanism of the host, gains footing in the tissues but is now exposed to a new set of conditions which are unfavourable to it. Finally, having overcome these obstacles, elimination from the body through excretion may temper the effects of the agent. In other words, the conflict resolves itself into four stages:

(1) At the time of entry
(2) After penetration of the host's outer defences.
(3) During spread throughout the host.
(4) At the time of exit from the host.

In each we can recognize measures of protection inherent to the component tissues of the host. Some of these are so efficient as to suggest an evolutionary process brought out through natural selection for the purpose of defence.

(I) Defence against penetration

Cells lining body surfaces and cavities are certainly concerned in preventing penetration from without, but they vary a great deal in effectiveness. Certain poisons, for instance, pass the barrier of the stomach epithelium more readily than others, some penetrate the skin without difficulty but others cannot get past this natural barrier so long as the skin is healthy. These factors are very clearly brought out when the disease-producing agent is itself a living organism, and it is well known that some bacteria penetrate the lining of the alimentary canal but are unable to pass the epidermis. *Bacillus typhosus* is such an organism. It is worth while considering the simple types of epithelium from this point of view.

(a) *Relative resistance of epithelia*

Spheroidal or cuboidal epithelium as found in the ducts of various glands is not very resistant to damage, whilst columnar epithelium is susceptible to mechanical and chemical upsets although less often injured by infectious agents. Squamous epithelium, which covers the skin and the communications with the exterior, is very resistant. This may be because it possesses remarkable powers of replacing its damaged and dead cells, a process which goes on under normal conditions and compensates for wear and tear. Sites of junction between squamous and other epithelia may be regarded as danger spots, especially when injured over a long period. Transitional epithelium, seen in the bladder, is resistant to mechanical damage and many common bacteria such as streptococci and staphylococci, but ciliated columnar epithelium is easily destroyed by irritants, bacteria and viruses such as those of influenza and the common cold.

Such resistance, however, may be broken down easily in certain places so that the agent penetrates the surface or cavity lined by epithelium. Transitional epithelium of the urethra or bladder, for instance, may be changed in this way by urine containing drugs such as formaldehyde or mercurials, or by the presence of a stone or a tumour. Bladder epithelium also shows remarkable differences in its behaviour to various infections, being fairly resistant to venereal micro-organisms but susceptible to tuberculosis. Stratified epithelium lining the fossa navicularis, para-urethral glands and crypts, or the oral mucosa, successfully presents a barrier to the gonococcus, the organism seldom penetrating beyond the second or third layer of cells where

phagocytosis generally occurs. Cylindrical epithelium lining the urethra, conjunctiva and rectum is quickly penetrated by the gonococcus. Ciliated epithelium of the Fallopian tubes and vasa deferentia is not so quickly penetrated possibly because of the movements of the cilia which pass the gonococcus onwards along these tubes. When this movement is interfered with, the organism quickly invades the epithelium and a gonococcal inflammation follows.

(b) Resistance varying with age of cell

It seems fairly certain also that the resistance of cells varies with their age. Some very young cells are more susceptible to damage than they are when older. Certain viruses, for example, vaccinia, herpes, influenza, can be grown upon the developing egg membranes of the chick but not in the adult tissues. The lack of differentiation or individuality of the cells of the membranes is thus associated with poor resistance to viruses.

The foetus is seldom the site of infection. No doubt this is partly due to the placental barrier interposed between the maternal and foetal circulation, and to its snug situation enclosed by a water cushion, but there is interesting experimental evidence which suggests that the foetus is for a long time insusceptible to bacteria. Foetal guinea-pigs of different ages injected with bacteria, ink or turpentine show a primitive histiocytic reaction only, even after a circulatory system has developed. Very young foetuses are anergic to inflammatory stimuli. When Vorwald (1937) introduced tubercle bacilli into foetal guinea-pigs he found that the bacilli grew very slowly and that there was merely a mononuclear-cell reaction without necrosis. Immediately after birth the reaction was similar to that in the early stages of foetal life but with increasing age it approximated to that of the adult. Vorwald explains this retardation in growth of the tubercle bacillus as being due to oxygen unsaturation of foetal blood—oxygen leaving the foetal heart is only 48 9 per cent saturated, a condition known to be unfavourable to cultivation of the bacilli upon artificial media. On the other hand, some organisms possess the power of invading the foetus by way of the placental circulation and destroying it. The spirochaete of syphilis is a well-known example.

Insusceptibility of foetus

The young child, too, has certain resistances and susceptibilities determined, it would seem, by:

(1) Inheritance, that is, its gene constitution.

(2) Transmission of antibodies from the mother by the placenta or during suckling. As evidence of the former we may instance the positive Wassermann reaction in the child of a syphilitic mother for a few weeks after birth, and of the latter Theobald Smith's proof of the transmission by way of the colostrum of antibodies against scours in calves. (Smith, Theobald and Little, 1922.)

Antibodies from mother

(3) Rate of growth of cells, perhaps; some cells grow so rapidly that damage is quickly made good by replacement. Newly formed cells or regenerated cells may be tolerant to certain poisons, for example, organic arsenicals, gold salts, uranium salts (MacNider, 1941, for reference to his previous work; Kuhs, Longley and Tatum, 1939; Bunting and Longley, 1940; Cortell and Richards, 1942). A change in epithelial type in the nose of the ferret may be associated with increased resistance to certain viruses (Francis and Stuart-Harris, 1938).

Some interesting examples may be mentioned. Guinea-pigs weighing less than 400 grammes react slightly and irregularly to diphtheria toxin injected into the skin. Tuberculous guinea-pigs less than one month old either do not respond to the tuberculin test or react slightly. Intracutaneous injection of virulent pneumococci into adult rabbits causes extensive local inflammation but seldom death; young rabbits fail to develop the local reaction though they succumb to bacteriaemia. Young children react very slightly to skin irritants such as turpentine, and possess poor defensive cellular mechanisms against streptococci.

Keratin barrier

Local variations also occur in the distribution of epithelium lining varicus parts of the body. The epidermis is much thicker in the sole of the foot and in the palm of the hand, especially in manual workers. The increased amount of keratin largely responsible for this thickening no doubt acts as a firm but flexible barrier to many injurious agents. Such anatomical changes may well represent adaptations to injury, a subject which will be touched upon later.

(c) *Special protective mechanisms*

Lysozyme

Special protective mechanisms are found in certain regions. Colebrook (1930) has described interesting experiments which suggest that the skin of the hands is capable of destroying various bacteria quickly, especially *Bacillus coli* and streptococci. The normal inhabitants of the skin are unaffected, in all probability because they are adapted to their environment. Colebrook thinks that this mechanism is connected with the sweat. In the tears Fleming (1922, 1929) discovered an enzyme, lysozyme, which quickly destroys some species of bacteria. Lysozyme has also been obtained from the skin and the alimentary canal. Mucous membranes secrete mucus which can trap and remove dust particles and bacteria. It is especially in the upper reaches of the respiratory apparatus that mucus exerts its useful action. At this spot there is a good deal of surface movement, provided by cilia attached to the epithelium, and it has been shown that ciliary movement is highly important in removing dust and bacteria and other foreign material from the lower levels of the respiratory tract (*see* Florey, Carleton and Wells, 1932, for discussion of this).

Ciliary movement

Mucous surfaces, too, are continually flushed by fluid. The mouth is bathed by saliva, the alimentary canal by various juices of diverse composition, and the movement of the muscles attached to these structures provides the propulsive power for flow. The arrangement of the tear-ducts ensures the carriage of injurious products, collected from the delicate conjunctival sac and cornea, to the exterior by way of the less delicate nasal passages.

Gastric juice

Some evidence also suggests a protective action on the part of the juices found in the alimentary canal. The acid of the gastric juice is to some extent antiseptic so that many bacteria entering with the food are killed in the stomach. The normal small intestine is free from bacteria.

Nasal lysozyme

The nose is certainly an effective barrier to infection, for its many compartments break up the air currents so that particulates are exposed to a considerable surface and trapped. Mucus is quickly secreted, often in large amounts, and assists in the removal of such particles. Nasal lysozyme destroys some bacteria. There is evidence, too, of highly efficient, though obscure, bactericidal mechanisms Thomson and Hewlett (1896), for instance, found that cultures of *Bacillus prodigiosus* placed on the nasal septum could

not be recovered from the nasal mucosa after two hours. *B. coli* suspensions sprayed on the nasal mucosa are destroyed in a few minutes. Repeated application of the organism does not exhaust this bactericidal mechanism. The paranasal sinuses, also, possess a similar protection. Calamida and Bertarelli (1902) showed that the healthy sinuses are bacteriologically sterile, and when organisms are introduced the sinuses become bacteria-free in 24 hours. The ciliated epithelium of the sinus removes foreign particles, but the olfactory mucosa seems to be a danger spot, for Rake (1937) has demonstrated the ease with which certain particles and viruses penetrate this region to reach the lymphatics of the cranial cavity.

Olfactory mucosa susceptible

In the external ear, valuable protection is afforded by the anatomical arrangement of the external auditory meatus, its hairs and waxy secretion. The eyes are guarded by the continual movements of the eyelids which aid the flushing action of the tears. The tonsillar rings of the nasopharynx, too, play an important part in resistance.

The vaginal mucous membrane is protected by its mucus and other secretions, whilst it possesses an interesting bacterial flora which controls the spread and growth of other bacterial species through the production of lactic acid. Due emphasis must be placed upon involuntary reflex movements such as sneezing, coughing, retching, vomiting and intestinal peristalsis in the removal of highly irritant substances from the respiratory passages and alimentary canal.

Involuntary reflex movements

(2) Defence after penetration of the host

Mechanisms of defence include the dilution and local precipitation of absorbed injurious agents, the immobilization and destruction of micro-organisms and the local production of specific antibodies against these. I shall return to this subject later.

(3) Prevention of generalized spread by removal from the blood stream

If the primary defence mechanisms are eluded the injurious agent may reach the blood stream in the first instance by way of the lymphatics (Noetzel, 1906; Barnes and Trueta, 1941), and be carried to the tissues of the host, reaching the vulnerable regions. This necessitates passage through the vessel walls and lymphatics, and here a further barrier of protection is often encountered in the lining endothelium of the vessels. Soluble poisons may be held up by adsorption at cell surfaces or taken up and dealt with in the cytoplasm. Molecular size determines the path of absorption of chemicals, smaller molecules entering the blood stream, the larger ones the lymphatics (Drinker and Field, 1933). Bacteria are ingested by endothelium modified for this function in certain regions of the vascular system, the so-called reticulo-endothelial system. Even when bacteria pass the endothelial barrier they soon encounter phagocytic cells in close association with the outer coats of blood-vessels. In the blood, too, there are many phagocytic cells, polymorphonuclear leucocytes and mononuclear cells, though it is uncertain whether phagocytosis goes on actively when these cells are in motion. Such cells rapidly increase in number under the stimulus of invasion. Furthermore, the efficiency of these mechanisms may be keyed up by previous contact with the injurious agents, the process being one of immunity. Thus pneumococci are

Lymphatic route of spread

Endothelial barrier

[Ph]agocytic [cel]ls

rapidly removed by the liver of an actively immunized animal but relatively few cocci are taken up by the normal liver. The pneumococci adhere to the Kupffer cells lining the smallest vessels of the organ. Living staphylococci and paratyphoid bacilli are quickly abstracted from the circulating blood in immune animals, less rapidly in normal, by various organs, especially the liver and spleen where they may accumulate in enormous numbers.

(4) Excretion and biological adaptations

Many harmful agents are dealt with by the ordinary channels of excretion. Poisons and toxins and sometimes bacteria are excreted in the bile, urine, sweat and faeces.

Biological adaptations

Finally, there is evidence of the development in the host repeatedly exposed to injurious agents of protective mechanisms of different types. These are best regarded as biological adaptations. Skin areas become thicker and harder (calloused) when frequently injured, as in certain occupations. The buccal mucosa is said to be thicker in glass-blowers. Nansen noted increased resistance to cold of exposed extremities in Arctic explorers. The prolapsed vagina and uterus become covered with more resistant keratinized epithelium. The guinea-pig's ear shows enormous epithelial thickening after repeated immersion in moderately hot water (56° C.) Exposure to strong sunlight is well known to increase the protective pigmentation of the skin through the mobilization of melanoblasts. The development of tolerance to drugs such as morphine or arsenic no doubt is to be included with these examples.

(5) Factors adversely affecting protective mechanisms

Fatigue

Little is known about the factors which adversely affect protective mechanisms, but there is reason to believe that fatigue may be important. Experimental animals subjected to exhaustive exercise in rotating drums and then inoculated with cultures of organisms die more rapidly or in larger numbers than non-fatigued animals similarly inoculated. Bailey (1929), for instance, introduced pneumococci into the nasal passages of rabbits subjected once or twice to strenuous exercise before or after the inoculation. The animals were less able to resist infection than normal controls. Their blood stream was more frequently invaded, the infection was more rapid and severe and a greater mortality resulted. The experiments of Boycott and Price-Jones (1926) with a natural infection of the rat gave similar results although no increase in susceptibility to human and bovine tubercle bacilli could be demonstrated. The experimental evidence would suggest that one or two periods of fatiguing muscular exercise following immediately after inoculation of animals with foreign organisms encourages infection. Human experience gives some support to this conclusion. Cowles (1918) recorded a higher incidence of pneumonia each year in schoolboys given to athletics than among the less athletic teachers or employees of the school. Pupils of a nearby school who limited themselves to everyday activities were also less affected. No cases of pneumonia occurred among pupils over a period of 5 years when the boys subject to, or convalescent from, mild respiratory infections were shielded from fatigue.

Vernon (1921) reported a close parallelism between the percentage time lost

from sickness among munition workers and the number of weekly hours of labour. Smelters, whose work is the most exhausting of all, lost 23 per cent more days from sickness than the average for all steel-workers, and showed a mortality of 26 per cent above the average for the whole group of steel-workers. Vernon emphasized the unfavourable temperature conditions, but considered that fatigue was probably a factor as well since diseases other than respiratory also showed some excess fatigue. The effect of muscular fatigue upon defence mechanisms of the body seems to be of a minor character. The permeability of mucous membranes for bacteria is thought to be increased by fatigue but there is some disagreement about this. Experiments on antibody content of blood after fatigue have also yielded conflicting results. Zlatogoroff and Kostereff (1931) found that following a march of 40 kilometres, the Schick and Dick reactions which, when negative, indicate the presence of antitoxin, became positive in a large group of men. Decreased ability to form typhoid agglutinins after exhaustive exercise has been reported by Lara and de Jesus (1930), Scalfati (1912) and de Sandro (1910). Bailey (1929) found increased production in exercised rabbits, but inconsistent results were recorded by Vallardi (1912) and Ficker (1906). Similar inconsistences have been noted with tests of the bactericidal power of the blood, and with haemolytic tests. A reduction of opsonic and phagocytic properties is reported in man and experimental animals. Ronzani (1907) claimed that the protective ability of the lung was decreased by fatigue. Fatigued guinea-pigs showed *B. prodigiosus* in lung cultures 72 hours after inhalation of organisms, whereas unexercised animals were able to eliminate all the bacteria by the end of 48 hours. Bailey (1925) found that 40 per cent of fatigued rabbits developed marked pulmonary lesions after intratracheal inoculation with type I pneumococcus as compared with 20 per cent in the controls. This was true even when fatigue preceded inoculation.

Antibody content of blood

Reduced opsonic and phagocytic properties

Little is known about the effect of fatigue on the reticulo-endothelial system, but a marked increase in the total leucocyte count has been recorded in tired people (Garrey and Butler, 1929). Lymphocytes increase first then neutrophils. Lymphopaenia follows.

Metabolic condition

There is some evidence, too, that the metabolic condition is of importance in resistance. Spaeth (1925) observed that guinea-pigs which showed a gain in weight following fatigue and infection invariably recovered. He concluded that resistance is a function of the anabolic rate after infection and showed that young fasting guinea-pigs and exhausted guinea-pigs, when fed immediately after inoculation with pneumococci, resisted the infection better than normally fed control animals.

Nutritional defects

There is strong evidence that the resistance of an animal to both spontaneous and induced infection is profoundly affected by nutritional defects, including generally restricted diets as well as those lacking particular vitamins (*see* Perla and Marmorston, 1941, for discussion of this). The normal irritability of the reticulo-endothelial tissue appears to be greatly reduced by defects of diet which lead to loss of weight.

Part played by glands

The glands of internal secretion, especially the adrenals, are closely connected with resistance to infection (Perla and Marmorston, 1933). A vast literature exists about the importance of vitamins and infection but this cannot be discussed here.

2. ACUTE INFLAMMATION

When injurious agents pass the surface barrier of the body and reach the underlying tissues they encounter a series of reactions in these tissues which we know as inflammation. Modern study has associated such responses with familiar clinical features which were recognized by the ancients and clearly defined by the Roman physician Celsus who lived in the Christian era. Though we agree that these features vary according to the situation and severity of the initial injury, we have little reason to discard Celsus's description of:

(1) Redness—rubor.
(2) Swelling—tumor.
(3) Heat—calor.
(4) Pain—dolor.

John Hunter (1792) added a fifth feature, diminution or loss of function. But clinical evidence of inflammation is by no means regular, for redness and heat are absent when avascular structures such as the cornea or hyaline cartilage are injured, pain may be slight when sensory nerves are few or have been put out of action, swelling may not be prominent in tissues with little powers of expansion, impaired function can be compensated for in organs with great powers of reserve, as, for example, in the liver and kidneys. Nevertheless, the response of the tissues to injury is astonishingly constant and there is little variation from region to region.

(a) *Reaction of the tissues to injury*

The simplest case is that of injury to an avascular tissue such as the cornea. Senftleben (1878), in a classical study, describes how he inflicted simple chemical injuries on this structure and introduced living bacteria into its tissues, thus grading the intensity of the injury. He also clarified the time sequence of events following each type of injury.

Slight damage of the surface epithelium by the application of silver nitrate or a caustic is followed by death of the injured cells and their rapid replacement through proliferation of the uninjured cells at the margins of the affected area. Newly formed corneal epithelium slides over the gap resulting from the detachment of the dead cells, thus re-lining the area. With a somewhat more severe injury the underlying corneal connective-tissue cells (corneal corpuscles) are also damaged. Within a short time the adjacent connective-tissue cells swell, divide and give rise to daughter cells which move into the damaged area and eventually restore deficiencies there. At the same time the destroyed surface epithelium is removed and replaced by new epithelium. When a severe injury of the cornea is produced by the cutting out of a piece, the cellularity of the surrounding corneal tissues increases by migration along the tissue spaces of small cells not normally found in this region. These cells are similar to the mononucleated cells in the blood and reach the cornea from the vessels of the conjunctiva. They accumulate round the dead tissue at the margins of the injured area and ingest it. The corneal corpuscles also divide, and the new connective-tissue cells so produced grow into and replace the gap, whilst the epithelium spreads over the bare surface. When bacteria such as staphylococci are introduced into the cornea by means of a fine needle they

Proliferation of uninjured cells

New surface epithelium

Ingestion of dead tissue

increase rapidly and spread some distance along the tissue spaces. Many polymorphonuclear leucocytes and mononuclear cells migrate from the blood-vessels of the conjunctiva and surrounding tissues towards the cocci and form clusters round the bacterial colonies. After a time some cells and cocci are destroyed. Pus corpuscles, which are nothing more than damaged or dead leucocytes, appear in increasing numbers and subsequently are discharged from the destroyed corneal surface. At the same time the conjunctiva

Reddening and swelling

becomes reddened and swollen; its blood-vessels are dilated and packed with blood cells and leucocytes can be traced from these vessels into the cornea. Whilst the cocci increase in number the process continues and grows in intensity until much of the cornea is affected. When further coccal growth ceases, migration of leucocytes from the conjunctiva tails off though mononuclear cells become more numerous. Normal corneal corpuscles at the

New connective-tissue cells

margins of the coccal growing area now divide actively, giving rise to new connective-tissue cells which replace those killed by the coccal invasion. Mononuclear cells seem to assist in this process by taking up debris of pus corpuscles, cocci and dead corneal cells. Eventually the corneal surface, injured during introduction of the bacteria and subsequently by bacterial growth, is covered over through proliferation and overgrowth of normal epithelium, recovery is complete and the reaction ceases. The conjunctival changes disappear soon after bacterial spread is checked.

These experiments bring out some of the most important facts about reaction to injury. In the first place, they show that injury destroys or alters the cells of a tissue. Provided a few cells only are affected, there is rapid replacement through division of the normal cells of the same type close to the site of injury. With severe injury the reaction is more pronounced and is no longer limited to the region injured. White blood cells migrate to the damaged area from a distance. Clearly these distant parts are influenced in some way by the injury. How this is done has been determined by a long series of experimental studies on inflammation in vascular structures, due to the pioneer efforts of Addison, Wharton Jones, Waller and Cohnheim.

(b) *Changes in vascular structure after injury*

The simplest method of studying changes after injury in a vascular structure is by means of a transparent tissue such as the frog's web, mesentery or tongue, all of which can be viewed for some time with the aid of a simple microscope and direct illumination. But the mesentery or omentum of mammals such as the rat, guinea-pig, rabbit or cat does equally well, although difficulties of prolonged anaesthesia make them much inferior to the frog. Provision must be made, of course, against drying and too severe injury. In recent years, Clark and his collaborators have developed a method for insert-

Microscopic and cinematographic observation

ing a transparent chamber in the rabbit's ear which allows of microscopic and cinematographic observation for long periods without the use of a[illegible]

si[illegible]

a regular series of changes is gone through in the vicinity [illegible]

Acceleration

Dilatation

flow of blood through the arterioles, capillaries and venules hurries up (acceleration) whilst the arterioles and venules widen (dilatation) Many

capillaries which were invisible before, open up and dilate. After a short time the blood stream begins to move more slowly until the red corpuscles may be seen as they pass by (retardation). In some vessels, the flow may even cease for a short time and then move on again, or may move backwards and forwards with jerky movements (oscillation). When the injury is severe, flow through vessels ceases completely (stasis) and clotting of blood is apt to occur then. But stasis, even though prolonged, may be followed by resumption of flow as the effects of injury wear off.

Retardation

Oscillation

Stasis

Whilst these changes are going on in the blood-vessels, fluid can be seen accumulating in the tissues round the site of injury so that the cells and fibres there appear well separated. Some of this fluid is quickly removed by the lymphatics, some contributes to the formation of delicate threads of fibrin, but much of it remains in the tissue spaces and may even bulge the surface layers outwards to form a bleb or weal. At the same time striking changes are going on within the blood-vessels. In normal arterioles and venules the cells are arranged in two more or less well-defined zones: (1) a central or axial zone in which nearly all the corpuscles, both red and white, rush along and (2) a peripheral or plasma zone against the vessel wall. When the blood flow slows down, the axial zone widens and corpuscles occupy the whole of the vessel lumen. Leucocytes approach the endothelium lining the venules (and somewhat later that of the capillaries), sometimes sticking to it and then being dislodged, finally becoming attached at one point, the rest of the body moving about in the blood stream. If such a leucocyte is watched closely a small projection of cytoplasm is seen to bulge outside the endothelial lining and this increases until eventually the whole cell appears to have passed through the vessel wall. Leucocytes go on migrating in this fashion and accumulate about the vessel or near it. Many pass towards the site of injury. Cohnheim believed that the leucocytes were forced out by the pressure of the rapidly moving stream but it is known from the researches of Wharton Jones (1850), Metchnikoff (1892) and many others that leucocytes are actively motile, and are attracted to areas of injury by chemical agents (chemiotaxis) produced there. Red corpuscles, too, may pass out in small numbers with the leucocytes (diapedesis). Sometimes these are so numerous as to suggest a haemorrhage.

Formation of a weal

Migration of leucocytes

Chemiotaxis

Diapedesis

The immediate responses to injury of a vascular tissue thus are:

(1) Vascular dilatation or local hyperaemia, the whole vascular bed widening out and the blood increasing in quantity in the affected area.

(2) Exudation of fluid, much of the fluid content of the vessels being poured into the injured tissues.

(3) Migration of cells and active movement of leucocytes towards the damaged areas.

It is obvious that these changes explain the clinical features of inflammation enunciated by Celsus and John Hunter.

(c) *Role of chemical secretions in inflammation*

There seems little doubt that the starting-point for inflammation is tissue death produced by the injurious agent. It is no exaggeration to say that all agents capable of killing tissue can produce acute inflammation and conversely all sorts of agents which induce inflammation can in stronger concentrations or intensities kill tissues. Moreover, such widely differing agents as

chemical and bacterial poisons, heat and cold, trauma, x-rays and radium produce the same sort of inflammation. Clearly, the only common factor must be tissue death. Inflammation proceeds in much the same fashion in tissues deprived of their nerve supply. MacCallum (1922) showed that if the limb of an animal were amputated and then replaced by anastomosis of the large vessels and suture of the other tissues so that all nerves were certainly divided, the application of mustard oil produced inflammation quite as pronounced in the denervated limb as in the normal one. The inflammatory changes appear after the same time interval and are, if anything, slightly more intense. The nervous system, therefore, cannot be the essential factor in producing inflammation. The possibility of a chemical agent associated with dead or damaged tissue has to be considered.

Tissue death the common factor

It has been shown that extracts of dead tissue contain a diffusible substance which is capable of inducing hyperaemia and exudation of fluids from vessels (wealing), two of the striking features of inflammation. Dale and Laidlaw (1919) obtained evidence from the study of traumatic shock that histamine, a derivative of the protein of injured tissues, may be responsible for dilatation of capillaries. Lewis, Grant and Marvin (1927) have given us much information supporting the view that histamine or some substance allied to histamine is responsible at least for some of the phenomena of inflammation.

Histamine

Histamine is the amine formed when carbon dioxide is split off from the amino acid, histidine, which is present in a variety of normal tissues; histamine has been extracted from normal ox liver and lung, spleen, heart and smooth muscle. Harris (1927) found that blood contains 0·3 milligram per kilogram, normal skin 10 milligrams per kilogram. Herbivorous animals have relatively large amounts in their lungs. Histidine is present in the intestinal mucosa whilst histamine-producing micro-organisms live in the human intestine. Histamine-like substances can be released from human skin by mechanical stimulation or by nerve impulses in sufficient amounts in certain individuals to cause flushing of the face, secretion of gastric juice or lowering of the blood-pressure similar to that occurring after the parenteral injection of histamine. Release of histamine occurs in guinea-pigs subjected to anaphylactic shock and it can be demonstrated both in the lymph and in the blood coming from the affected organs. Snake venoms and bacterial toxins liberate histamine from the perfused lungs of the guinea-pig and cat. Normal mechanisms for controlling the action of histamine have been suggested. Thus many physiological processes are affected in opposite directions by histamine and epinephrine whilst epinephrine is liberated from the adrenal glands after injection of histamine. Hence these substances may be physiological antagonists. There is evidence, too, of a specific enzyme for the inactivation of histamine in certain tissues (histaminase).

Histaminase

Lewis and his school have shown that the intradermal injection of very small amounts of histamine phosphate produces effects similar to those following many forms of mechanical or chemical stimuli. This reaction has been termed the "triple response" and includes:

The "triple response"

(1) Vasodilatation of the small vessels at the site of injection.

(2) A more diffuse "flare" extending for 3 or more centimetres.

(3) Wealing, or an area of local oedema which appears soon after the flare and extends over a similar area. This effect usually disappears within an hour.

The weal develops even when the nerves to the skin have been cut, but not the flare, so that the former is independent of the nervous system; the latter apparently depends upon the integrity of the axons of sensory nerves from the subcutaneous tissue (Lewis, Grant and Marvin, 1927). The flare is also abolished by application of local anaesthetics such as cocaine or Novocain. Wealing is a manifestation of increased permeability of vessels for plasma.

Local anaesthesia

Lewis (1927) suggests that the response to tissue damage is caused by a diffusible substance resembling histamine which he prefers to call "H" substance. The action of "H" substance is very prolonged in contrast to the short response evoked by histamine, so that possibly other products are also concerned. Krogh (1929) believes that there is more than one chemical substance responsible for the triple response and advocates the term "H" colloid to indicate the possibility of a less diffusible vasodilator.

"H" substance

"H" colloid

It seems very likely that acetylcholine is responsible for the flare.

Acetylcholine

In recent years Menkin (1937) claims to have obtained from injured tissue a diffusible, crystalline substance, probably a polypeptide, designated leukotaxine, differing from histamine. Minute amounts of this factor induce considerable exudation from capillaries of plasma proteins which coagulate and according to Menkin form obstructive thrombi in the afferent lymphatics, so inhibiting the spread of the irritant. Leukotaxine also encourages migration of leucocytes. Histamine fails to do this.

Leukotaxine

Duthie and Chain (1939) and Cullumbine and Rydon (1946) have also obtained a polypeptide from peptic hydrolysates of blood fibrin, which makes the blood-vessels more permeable to plasma and encourages leucocytic infiltration when it is injected into the skin of rabbits and guinea-pigs. It exerts a strong chemiotactic action on leucocytes *in vitro*. Polypeptides with similar properties are found in peptic hydrolysates of serum albumin and globulin, casein, egg albumen but not gelatin. Such compounds would account for the chemiotactic action of damaged tissue as well as of pure proteins, and of micro-organisms such as the staphylococcus (McCutcheon and Dixon, 1936).

Polypeptides

It is possible, therefore, to express the essentials of acute inflammation in terms of response to chemical substances produced by the action of the injurious agents upon the affected tissues. The vascular dilatation and exudation of fluid (triple response of Lewis) are the result of histamine or "H" substances, and Menkin's polypeptide acting upon the capillaries and acetylcholine acting upon the arterioles. Migration of leucocytes is induced by some product of tissue death which is quite likely leukotaxine. There is evidence, too, that bacteria or bacterial products can attract leucocytes (Ziegler, 1901; Marchand, 1921) whilst leucocytic constituents also possess similar properties. *Staphylococcus albus* is especially potent as a chemiotactic agent.

(d) Role of the nervous system

Although inflammation can proceed as usual in tissues deprived of their nerve supply (hot-water bottle burns are the same in patients with transverse lesions of the spinal cord as in normal individuals) there is reason to believe that the nervous system can modify the inflammatory response. In a well-known experiment, Samuel (1890) divided the cervical sympathetic nerves to one ear of a rabbit and the parasympathetic (auricular) nerves to the other

ear. Vasodilatation developed in the one case, vasoconstriction in the other. Both ears were then plunged into water at 50° C. The first showed severe inflammation, the second responded very slowly and developed necrosis rather than inflammation. There is evidence, too, that denervated organs are more susceptible to poisons than are intact organs. It is said that vascular poisons such as snake venoms cause greater injury in the denervated kidney than in the normal organ.

(e) *The nature of inflammation*

Protective mechanism

It is tempting to regard inflammation as a protective mechanism, especially as injurious substances become localized or fixed at the inflamed site and are hindered from spreading through the host. It has been said that inflammation protects the organism as a whole at the expense of local injury (Menkin, 1940). Proof of this fixation mechanism depends upon observations such as the following. (1) Bacteria and inert substances—dyes, inorganic salts, graphite particles, foreign proteins like egg-white—introduced directly into inflamed tissues are fixed there (Burrows, 1932; Menkin, 1940) Peritoneal injection of sterile irritants (broth, saline) temporarily increases resistance to a subsequent injection of bacteria by setting up a mild peritonitis. Beef broth introduced into the skin of rabbits protects against staphylococci inoculated there 24 hours later. Aleuronat peritonitis prevents blood dissemination of haemolytic streptococci (Opie, 1924). (2) Foreign substances and bacteria circulating in the blood stream localize in inflamed areas. It has long been suspected that injury may determine the localization of bacteria or viruses in damaged tissues—*locus minoris resistentiae*. Tubercle bacilli, for example, leave the blood stream to localize in subcutaneous lesions produced by injected silica (Kettle, 1927). Spirochaetes of syphilis introduced into rabbits by testicular inoculation or intravenously localize in distant skin wounds (Chesney, Turner and Halley, 1928). Streptococci lodge at the site of sterile abscesses, staphylococci in injured bone or where circulation is sluggish, such as the metaphysis of bone. Dyes, iron salts, foreign proteins, horse serum and graphite after intravenous injection become concentrated in inflamed areas (*see* Menkin, 1940, for literature).

Localization of bacteria

Several explanations of localization have been suggested. Opie (1924) and Menkin (1930) stress the "walling off" of the inflamed area through enhanced passage of fibrinogen from more permeable capillaries, and the resulting formation of a fibrinous barrier against absorption. Thrombi occlude the lumina of draining lymphatics so that there is a kind of blockade. Menkin places much emphasis upon this early fixation in a severely injured area for it ensures an interval in which the relatively sluggish leucocytes may assemble. Fibrin also constitutes a framework on which the phagocytes move or become anchored whilst they ingest their meals. Rich (1936), however, stresses physico-chemical factors in localization. Blood proteins can adsorb colloids and so may contribute to fixation. Other factors are known which exert an influence on the localization or spread of foreign substances in the tissues. Duran-Reynals (1929) and McClean (1930) described a "diffusing factor" in testicular extract which promotes the spread of bacteria and inert substances when introduced with these into tissues. McClean (1936) has also shown that certain bacteria exert a similar effect upon tissue permeability. Duthie and

Fibrinous barrier

[Ph]ysico-[ch]emical [fac]tors

Chain (1939) have given reasons for believing the diffusing factor to be hyaluronidase, an enzyme which promotes softening or lysis of tissue mucins and thus allows of diffusion. But mechanical factors are of minor importance when bacteria are susceptible to the action of leucocytes (Clark, 1929; Cannon and Hartley, 1938) or macrophages (Gay and Morrison, 1923). The barrier is of value chiefly in the defence against micro-organisms for which the body possesses an imperfect resistance, temporarily localizing these invaders until they can be attacked by the phagocytes (Rivers and Tillett, 1925; Rich, 1930). It is also possible that altered local conditions, such as change in reaction of the exudate, may play a part. Local acidosis is said to modify leucocytic responses (Menkin, 1940).

Hyaluronidase

Local acidosis

3. MODIFIED INFLAMMATORY REACTIONS

The fundamental inflammatory reaction, as discussed above, may be modified by differences in the injurious agents as well as by host variations.

Kettle (1927) has drawn attention to the "partiality of certain organisms for certain tissues". The staphylococcus commonly infects the skin and bones, the pneumococcus the lungs, the diphtheria bacillus makes for the upper respiratory passages, the typhoid bacillus the lymph follicles of the small intestine. The nature of this partiality is not known. Trauma may be important in some instances as in the localization of the gas gangrene organisms in muscle. A few examples of modification are worth while considering.

(1) Pyogenic cocci

In general, staphylococcal lesions are more restricted than streptococcal, the organisms forming colonies in the centre of a massive accumulation of leucocytes. Many leucocytes die at an early stage and undergo liquefaction, hence pus is soon produced and an abscess results. Streptococci are apt to affect mucous membranes and give less localized lesions, the organisms spreading widely through the tissues, especially in the lymphatics, so that the whole process is often more rapid and extensive. In erysipelas, for example, the tissues become infiltrated with streptococci and much fluid exudate forms in the looser areas whilst mononuclear cells rather than polymorphonuclear leucocytes accumulate round the organisms. Indeed, the streptococcus possesses a leucocidin which destroys leucocytes and dissolves fibrin.

Abscess formation has been studied intensively by many workers, but the early description by Hohnfeldt (1888) has seldom been surpassed and is well worth a detailed account. Four hours after staphylococci enter the subcutaneous tissues, cocci can be traced between the connective-tissue lamellae, and are present in connective-tissue cells (probably histiocytes) and leucocytes. The latter are mostly mononuclear except along the line of infection where a few polymorphonuclear cells are found. Adjoining blood-vessels are congested and margination of leucocytes can be seen.

Abscess formation

After 10 hours, the cocci are chiefly in the tissue spaces but also within leucocytes and fixed connective-tissue cells. Margination and migration of polymorphs are going on in the vessels, resulting in cellular infiltration of perivascular lymphatics and tissue spaces. Polymorphonuclear leucocytes are grouped round cocci with mononuclear cells in the coccus-free margin.

Cellular infiltration

After 20 hours, the cocci and leucocytes have increased but as yet there is no actual abscess. Connective-tissue fibrillae are pressed together and their nuclei obscured by leucocytes.

After 48 hours there is a sharply defined abscess composed of colonies of cocci closely surrounded by masses of polymorphs. The cocci decrease in number from the centre to the periphery where they are scanty. There are no new mononuclear cells at the periphery and mitoses are not seen. Connective-tissue cells are degenerating at the centre of the abscess and blood-vessels have completely disappeared there. At the periphery vessels are congested and show margination and migration of amoeboid leucocytes. Cocci are present in great numbers within the emigrated leucocytes, but show no alteration in form or staining. At the periphery they are few in number in the intercellular spaces and the fixed connective-tissue cells. They can also be demonstrated within the endothelium of blood-vessels, especially when migration is going on.

Migration of amoeboid leucocytes

After 4 days the centre of the abscess shows nuclear remnants, degenerated leucocytes and fragments of connective tissue; the periphery is a mass of polymorphs in the tissue spaces. Cocci appear to be proliferating within the abscess and they invade and destroy leucocytes without apparently undergoing much damage themselves. There is little evidence of new formation of connective tissue at the periphery.

By 8 days there is a well-defined abscess cavity containing pus but no pronounced reaction in the fixed connective-tissue cells at the periphery.

Rupture or encapsulation

After 10 days there is rupture of the abscess or encapsulation of thickened pus. Coccal masses appear in the detritus. The capsule consists of granulation tissue with numerous capillaries, newly formed connective-tissue cells and large mononuclear cells. Evidence of division of cells is clear.

(2) Pneumococcus

The most characteristic feature of pneumococcal inflammation is the exudation of a fluid rich in proteins. This coagulates to form a thick meshwork of fibrin. The subsequent course of the infection follows the usual plan.

(3) Gonococcus and meningococcus

Phagocytosis of these organisms by the cells of the inflammatory exudate is very striking and the cocci actually proliferate within the phagocytes. It *would almost seem that phagocytosis is less of a defensive mechanism than* an aid to infection, and Kettle suggests that this phenomenon may explain how meningococci pass from the throat to the meninges and how gonococci reach the submucous tissue of the urethra to set up chronic inflammation. (*It may be said, in parentheses, that a great deal of fluid exudation in inflam*mation with the consequent increase in lymph flow will facilitate bacterial dissemination (Rhoads and Goodner, 1931; Rich and McKee, 1934; Lurie, 1936).)

(4) Diphtheria bacillus

This organism, through liberating an exotoxin, produces necrosis of the mucous epithelium, especially of the throat, with an intense vascular reaction and outpouring of large quantities of coagulable exudate. In the fully developed lesion the mucosa is entirely necrotic and replaced by a thick "false

membrane" of coagulated exudate and polymorphs. The bacilli are located on the surface of the membrane and do not penetrate to the deeper tissues. Beneath the membrane there is a severe inflammatory reaction.

(5) Anthrax bacillus

The vascular reaction is pronounced and there is an abundant accumulation of leucocytes, and much exudation of fluid which passes into the epithelium and causes the typical ring of blisters (Kettle, 1927). The fluid does not coagulate easily, hence fibrin is not a characteristic feature of anthrax lesions. Bacilli are numerous but phagocytosis is seldom prominent except in the older parts of the lesion.

(6) Anaerobic bacilli

These organisms (*Clostridium welchii, Bacillus oedematiens, Vibrion septique*—the bacilli of gas gangrene) grow best in deep wounds especially when muscle is involved. The characteristic change produced is necrosis accompanied by gas formation. There is little or no inflammatory reaction. A serous exudate may form, but there are no inflammatory cells except in the cutaneous and subcutaneous tissues where many polymorphs assemble.

(7) Typhoid and dysentery bacilli

Typhoid bacilli attack the lymphoid tissue of the intestine, the spleen, mesenteric glands, skin, gall-bladder and bones Though an acute disease results, the polymorphonuclear leucocytes take little or no part in the inflammatory reaction, macrophages being mainly concerned.

Dysentery bacilli produce a diphtheroid lesion with widespread ulceration and destruction of the mucous membrane of the intestine in which both leucocytes and mononuclear cells are involved.

4. PHAGOCYTOSIS

We have seen that a great variety of mechanisms exist within the host which can be regarded as protective in nature. Perhaps the most outstanding of these is the property possessed by many cells of taking up and immobilizing material foreign to the organism. The term phagocyte is reserved for cells which are concerned mainly, if not wholly, with this function, especially the ingestion of bacteria, dye particles, dust, damaged cells and tissue debris. It is well to remember, however, that all cells possess the power of ingestion though in most cases this is limited to matter in solution. But there seems little doubt that even these cells can on occasions take up particulate matter and store it. Various kinds of epithelium, for instance, may become phagocytic for red blood cells or foreign particles. In the restricted sense of pathology and comparative physiology, phagocytes are cells which can ingest and digest particulates and have the power of independent movement by which they approach their prey. Many phagocytes pass through a phase during which they settle quietly in certain places, especially close to blood-vessels. With appropriate stimuli, however, they again become mobile and move towards a focus of attraction. The most potent attraction is an area in which inflammation is going on, and the evidence we have already discussed suggests that chemical substances are produced in such foci which are responsible for attraction of phagocytes.

Static phase

The Russian biologist Elie Metchnikoff (1892) recognized the existence of a class of cells mainly devoted to phagocytosis. He pointed out that in the lowest forms of life the cell is capable of taking up all sorts of material, whether soluble or particulate. The amoeba flows round particles of slime, clumps of bacteria or pieces of grit, incorporating them in its cytoplasm. Indigestible material is soon rejected, digestible stuff is enclosed within a vacuole and dissolved by enzymes. Chemical attraction (chemiotaxis) exists, for the amoeba moves towards some substances and away from others. Protozoa-like myxomycetes are well known to be attracted by certain plant infusions such as oak bark (Stahl, 1884). How far such movement is vital and purposive and how far the result of interplay of surface forces is not yet decided. Jennings (1902), for instance, has pointed out that many of these features can be mimicked in models. A drop of clove oil in a mixture of alcohol and glycerin flows towards a region where the alcohol is stronger or has been heated. A drop of chloroform in water engulfs particles of shellac and rejects wood or glass. It will coil up threads in much the same way as the amoeba coils up filaments. Too much emphasis must not be placed upon these analogies, but they serve to illustrate the simplicity of some of the phenomena.

Part played by surface forces

Amongst all forms of life the phenomenon of phagocytosis is well established. In vertebrates, phagocytes fall into two main classes, (1) wandering and (2) fixed. The former include the blood phagocytes and the wandering tissue cells. A great deal of specialization is found in the blood cells and we recognize two important types, (1) polymorphonuclear leucocytes and (2) macrophages. Of these, the polymorphonuclear cells are the most prominent in the early stages of all forms of inflammation and throughout the course of acute inflammation. They soon migrate from blood-vessels to areas of damage, and at once engage in phagocytosis so that they are held to be the most active phagocytes. Actually, there is little difference in phagocytic properties between polymorphs and macrophages, but the polymorphonuclear cells being more numerous in the blood are more readily available, whilst they move more rapidly than other cells so that they reach the injured area sooner. They are also mobilized from the bone marrow into the blood stream with speed (leucocytosis). In contrast, many macrophages are formed locally, a process which takes time.

Polymorphonuclear leucocytes and macrophages

Other blood cells possess some degree of phagocytic power. Lymphocytes, the most mysterious of the blood cells, are probably connected in some way with toxin destruction rather than with phagocytosis although they are numerous in certain infections, especially syphilis. Each day lymphocytes migrate from the blood and lymph into the alimentary canal and Bunting and Huston (1921) suggest that this has to do with toxin destruction in that locality. Glimstedt (1936) has succeeded in keeping young guinea-pigs free from bacteria for 60 days after birth and notes in such animals the poor development of lymphoid tissue. Myeloblasts are actively phagocytic, myelocytes only slightly so. Eosinophilic and basophilic leucocytes differ in no wise from neutrophilic cells. Megakaryocytes are phagocytic for cells.

Toxin destruction by lymphocytes

The fixed phagocytic cells are conveniently grouped together as the reticulo-endothelial system (Aschoff, 1924). Broadly speaking, it includes collections of fixed and wandering cells disposed about the vascular system of the body. In certain situations, notably the liver, spleen, lungs, bone marrow and

Reticulo-endothelial system

lymphoid tissue, the reticulo-endothelial cells are especially numerous, constituting a highly labile clearing mechanism for the blood. All of these cells share the common properties of phagocytosis, motility at some stage in their history, the power of producing reticulum, a precursor of collagen which is important in repair of tissues, and response to chemiotactic agents. But it must be emphasized that even in the higher vertebrates many types of cells can act as phagocytes. Graff (1927) noted that in mushroom poisoning, for instance, hepatic cells are often filled with red corpuscles, the Kupffer cells frequently remaining empty. Rössle (1907) found red corpuscles in the epithelium of the liver, pancreas and kidneys in cases of haemochromatosis. Carleton (1931) showed that the vaginal epithelium can take up bacteria and foreign particles such as carbon. Conjunctival epithelium will ingest gonococci. Even fibroblasts are slightly phagocytic. Obviously the definition of phagocyte should not be too rigidly enforced.

Humoral factors

Finally, it is well recognized that other factors, which we call humoral, play an important part in protection. These are in solution in the tissue fluids and blood and probably are products of reticulo-endothelial and lymph-node activity. The existence of such humoral factors has been demonstrated in lower forms of life, for example, insects and fishes, and they assume increasing importance with the evolution of species. They are thus found at the sites of production of phagocytes and are closely mixed up with phagocytosis, to which they serve as adjuncts. Some humoral factors (opsonins) act directly upon bacteria and make them more suitable for phagocytic action There is considerable evidence that active immunization enhances the phagocytic activity of polymorphonuclear leucocytes and macrophages. This has been demonstrated in connexion with the pneumococcus (Wright, 1927), staphylococcus and paratyphoid bacillus (Cannon, Sullivan and Neckermann, 1932) and the streptococcus (Tsuda, 1923). Bloom (1927) showed that the phagocytosis of red cells by macrophages in tissue culture is greatly increased by the addition of specific immune serum. The efficiency with which bacteria are removed from the blood by the liver and spleen is greatly increased by active immunization of the animal (Manwaring and Coe, 1916).

Opsonins

Phagocytosis is thus one of the oldest protective mechanisms possessed by the organism against infection; it cannot be dissociated from the medium in which it takes place, nor does it seem likely that humoral mechanisms of defence reach their greatest efficiency without the intervention of cells.

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INJURY—CIVIL AND INDUSTRIAL

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1. INTRODUCTION

193.] This century has witnessed the establishment of various surgical specialties brought about by such advances in surgical technique as to demand the full attention of those who aim to master their subject. Such specialties as orthopaedic surgery, plastic and facio-maxillary surgery, the surgery of the peripheral and central nervous systems and thoracic surgery have made substantial contributions to advances in the treatment of accidental injuries. The surgery of accidents is not so much a new specialty in its own right as an attempt to organize and co-ordinate all modern advances in surgical treatment, so that they may be made immediately available to those injured by accident.

The need to develop modern facilities for the treatment of accidents was defined in *The Final Report of an Inter-Departmental Committee on the Rehabilitation of Persons Injured by Accidents* (1939).

Some of the conclusions and recommendations of this Committee were:

(1) "The total number of injuries by accidents of all kinds treated yearly in the hospitals of Great Britain is in excess of 1,250,000, of which fractures account for 200,000. These figures represent an enormous loss in working-time, production, wages, compensation and sickness benefit in many other ways."

(2) "The system which was universal till a comparatively recent date and still prevails to a large extent in the hospitals of Great Britain, by which fractures and other accidents are treated in the general surgical wards and under general surgical routine, is gravely defective and a radical change of method is necessary."

(3) Although "directed to enquire into the arrangements for the treatment of persons injured by accidents", they "considered that this was a wide field

and did not attempt to explore the whole of it". They outlined a scheme for the development of fracture services throughout Great Britain and contented themselves by suggesting "the possibility of developing this service into a more general accident service".

In one of the oldest known Egyptian records 48 surgical cases, of which 45 were due to trauma, are described in detail. Since this record of 5,000 years ago surgical literature on the treatment of accidents has been extensive, yet even in such company the Inter-Departmental Committee's Report is of historic note, for it is the first record of a Government's serious consideration of a surgical problem affecting the social, working and economic lives of a peace-time community.

The Report indicates that the reorganization which they recommend would be extensive in scope as well as radical in detail. The old system provided hospital treatment for 1,303,478 patients in 1,318 hospitals in 1935. Great though this total is, it does not take into account other sources of accident treatment. Outside the hospital services, private practitioners and the industrial medical services treat the more trivial cases, but these constitute a greater number of accidents each year than are treated by hospitals. Factories alone employ 200 full-time and 900 part-time medical officers and a nursing service of 9,000; of the latter group, 5,000 are State-registered (Stewart, 1946). This service alone treats a great number of accidents on the factory premises, rendering it unnecessary for the injured workpeople to leave their jobs Further, through its medical and welfare services, industry has shouldered a large measure of responsibility in the final rehabilitation and the resettlement of its workpeople back in work after hospital treatment.

The conclusions of the Inter-Departmental Committee's Report place the responsibility for accident treatment, rehabilitation and resettlement squarely upon the hospital service of Great Britain. Whilst this responsibility must in large measure be accepted, it is obvious that hospitals must seek to co-ordinate their work with other bodies, among which industry and its medical and welfare services are the most important.

2. SOCIAL CONSIDERATIONS

Analysis of accident figures

An analysis of accident figures taken from national hospital records shows that 55 per cent of accidents occur during normal domestic and recreational activities, 30 per cent occur at work and 15 per cent are the result of road traffic accidents. Patients represent a fair cross-section of the community and include men, women and children (Inter-Departmental Committee's Report, 1939).

Hospital experience indicates that children below the age of 5 years are particularly prone to accidents from burns and scalds in the home. From the age of 5 years children begin to take risks out of doors, and have little fear and no sense of safety in their quest for knowledge or adventure. They suffer a wide variety of injuries in their many pursuits. Happily they have a remarkable power of recovery from both bone and soft-tissue injuries.

Youth's characteristic curiosity continues during the first 2 years of employment, the accident rate for juveniles in industry being 10–15 per cent higher than in the adult worker. In the late teens and the early twenties youth has a

greater regard for road speed than for road safety, and the most serious road traffic accidents occur in this age-period in motor-cycleriders. At work, certain injuries are characteristic of various types of employment, for example, crush injuries of the spine, chest, pelvis and abdomen are most frequently encountered in miners and workers in heavy industry, fractures of the spine and the os calcis in builders and window cleaners, injuries to the hands in mass-production engineering and assembly work and industrial dermatitis in chemical workers.

From the age of 45 years onwards, accidents as the result of falls within the home become common in women. It is interesting to note how the incidence of fractures around the wrist changes with age and with sex as the years advance. In the age group 10–25 years, fractures in and around the wrist occur more frequently in males, but in the age group 45–65 years onwards women outnumber men with these fractures in the ratio of 5:1. As age further advances muscles and joints lose their elasticity, balance becomes less steady, falls are more frequent and lower limbs are fractured, again more commonly in women. These fractures occur particularly in and around the femoral neck. The solution of this problem calls for an unexpectedly high proportion of hospital beds for elderly female patients.

3. SURGICAL CONSIDERATIONS

The over-all surgical possibilities in the treatment of accidents are substantial. If modern advances in surgical practice can be made immediately available to all civilians who sustain injuries, then the death rate from accidents (now over 10,000 each year) can be lessened, permanent physical disabilities can be minimized or prevented, and the period of treatment necessary for any injury shortened. The immediate task is to change such a surgical possibility into a practical reality.

The first essential is to train surgeons with a general knowledge of the surgical problems presented by injuries resulting from accidents. That an accident service could develop naturally from the fracture service, which is suggested by the Inter-Departmental Committee, is a remote possibility; a much more positive plan is necessary to expedite such a development.

Surgical requirements

The major surgical requirements of an efficient peace-time accident service may be summarized.

(1) Treatment of traumatic shock

The first essential is to base shock treatment upon an early and accurate diagnosis of the injury or injuries requiring urgent surgical treatment. The aim is to resuscitate a patient until he is in a fit state to undergo essential surgery and, not infrequently, to assess priorities in treatment to the victim of multiple injuries. For example, in a shocked patient who is suffering from a crush fracture of the spine and a ruptured spleen, shock treatment is aimed at resuscitating him until he is fit to undergo splenectomy, the treatment of the spine taking second place.

Obviously all facilities for the treatment of shock must be immediately available in an accident hospital. Yet it is no longer sufficient to bring the very gravely injured patient into hospital before adequate shock treatment is commenced. The Birmingham Accident Hospital was recently asked to receive a

very badly injured patient, immediately after a car smash, from a cottage hospital 40 miles away. The injuries were obviously very severe, and the journey was long. The Hospital sent its mobile surgical unit (Fig. 35) with surgical staff to the patient. His injuries included a severe compound mid-third face fracture with the loss of one eye, fractures of the femur, patella, humerus, clavicle, both bones of the forearm and a complete dislocation of the knee. At the cottage hospital the following shock treatment was commenced: immobilization of the multiple fractures, blood transfusion and measures for the relief of pain. Resuscitation by oxygen inhalation and blood transfusion was continued during the journey back (Fig. 36). The patient arrived in hospital in excellent shape and withstood all subsequent plenary surgical treatment.

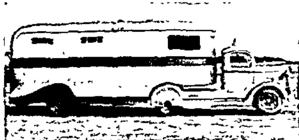


Fig. 35.—Mobile Surgical Unit.

Only such treatment could maintain a badly injured patient's general



Fig. 36.—Shock treatment with oxygen inhalation, blood transfusion and fracture immobilization carried out during transportation to hospital.

condition while he is being transported to a hospital at which facilities are adequate for the treatment of his many injuries.

(2) Treatment of open wounds with skin loss

Open wounds with skin loss are a frequent result of civilian accidents if all wounds are considered irrespective of their size, and particularly is this



FIG 37.—Extent of skin loss shown Plate fixation of fracture. Cross-leg skin flap being placed in position.



FIG 38.—Healed state of right leg Medial view.



FIG. 39.—Posterior view of both legs 12 months after injury. Note absence of oedema in both ankles.

ipplicable to finger and hand injuries. In civilian practice such wounds are presented for treatment within an hour of injury and, if trained surgical staff are available, may be safely closed by modern methods of skin grafting after careful excision. The whole procedure is made safe by chemotherapy and penicillin. The extent to which this technique may be carried is illustrated by the following case.

Chemotherapy and penicillin

12.8 46. F.T., male, age 28.

Right leg trapped under a heavy machine at work Compound fracture of both bones with full-thickness skin loss 6½ inches × 5 inches

Procedure Dead skin and muscle excision, wound toilet. Tibia reduced and immobilized by vitallium plate and screws, wound closed by cross-leg flap (Fig. 37) and split-skin grafting Donor area closed by split-skin graft. The two legs immobilized in a uniting plaster cast.

6 9 46 Cross-leg flap divided and base sutured into position on donor leg. Some raw granulating areas covered by split-skin graft.

10.9 46 All wounds on right leg soundly healed (Figs. 38 and 39) Donor leg healing

19 10 46. Discharged from in-patient treatment in plaster and on crutches.

14 11 46 Plaster removed, right leg supported in walking caliper, fracture clinically united.

Accident surgeons must be well trained in the technique of skin grafting. It is unlikely that in the near future the routine treatment of open wounds with skin loss will follow the principles of Winnett Orr (1929) or of Trueta (1939), at least in the civilian type of injury. It is still wiser to delay closure of wounds caused by the explosive violence so common in war, complicated as such wounds frequently are by serious vascular damage and by the risk of anaerobic infection.

(3) Treatment of burning and scalding accidents

Advances in the treatment of burning and scalding accidents, stimulated particularly by Colebrook (1947) in Great Britain, have demonstrated the absolute need of segregating these injuries in special wards equipped with special facilities, if treatment of such large open wounds is to be adequate.

The aims of treatment in these cases are:

(a) To prevent the onset of serious shock by means of controlled plasma transfusion.

(b) To prevent infection and cross-infection by the most rigid dressing technique in a dust-free atmosphere.

(c) To prevent and to treat nutritional disorders in the severely burned or scalded patient.

(d) To make good a full-thickness skin loss by very early skin grafting.

(e) In the early stages of treatment, to exercise especial care in preventing deformities, by means of adequate splintage in positions of function, early muscle and joint reduction and the use of saline baths.

(f) Finally, when the healed stage is reached, to have highly skilled plastic surgeons readily available to minimize the cosmetic and functional skin deformities of healed scars.

(4) Treatment of hand injuries and hand infections

Hand injuries and hand infections are an extremely common result of factory accidents By the application of advances in surgical technique pioneered by Bunnell (1946) many of the permanent disabilities which commonly result

from hand injuries, can be minimized. The more advanced of these techniques

Flexor tendon suture

—for example, flexor tendon suture in fingers—demand considerable and continued practice, and the surgical assessment of a severe hand injury requires considerable experience. If the surgery of the hand is to advance, then the treatment of these injuries must not be the responsibility of junior, inexperienced resident staff.

In the treatment of finger and hand infections an early and accurate diagnosis can entirely eliminate the crippling hand deformities not uncommonly seen a decade ago.

Closed hand infections are almost invariably presented for treatment *at an early stage* as either pulp or tendon-sheath infections, and if they receive adequate surgical treatment at that stage the really crippling fascial-space infections do not occur.

Surgery by efficient drainage is the method of choice in the treatment of pulp infections. The surgery of tendon-sheath infections has radically changed; the operative drainage of the sheath is now minimal but the diagnosis must be made early and the *systemic* use of penicillin must be adequate.

(5) Treatment of fractures, dislocations and sprains of the extremities

These injuries to the locomotor system form the largest single group of major injuries resulting from accident. Ninety per cent of such injuries are uncomplicated and, although their immediate surgical treatment calls for a high degree of manipulative and sometimes of operative skill, their early surgical treatment presents no real difficulties. It is, however, in the careful after-care of these injuries that special facilities are essential—the supervision of a limb immobilized in plaster, the early, continuous, well-planned and supervised programme of rehabilitation to prevent muscle wasting, and the development of preventable deformity.

Complications

In half the number of fractures of the extremities with complications, the complication arises out of the fact that the injury is associated with a communicating skin wound. In others the fracture is but one of several injuries and may or may not be the most severe injury.

It is around the problem of "associated injuries" particularly to the brain, the thoracic contents or the abdominal and pelvic viscera, that the modern tendency to establish accident surgery as a new specialty within medicine is most attacked and is most open to criticism.

(6) Treatment of associated injuries

Injuries to the head and the chest are the most important since they are the most frequent of the associated injuries.

At this stage in the development of cerebral and thoracic surgery there are not enough specialists available to undertake the treatment of all cerebral or all thoracic injuries. The solution of this problem would seem to be the establishment, within an accident service, of one special unit for cerebral injuries and another for thoracic injuries, under the supervision of acknowledged specialists upon whom would rest the responsibility for the training of general accident surgeons in the traumatic surgery of cerebral and thoracic injuries. The need for close team-work in the treatment of the victim of multiple injuries is very obvious. Sometimes the urgency of treatment of a

cerebral or thoracic injury dominates the whole clinical picture; in other cases these injuries must take second place to more serious matters. Various surgical specialties have neatly subdivided the human body into regions of specialized operative procedure but the problem of treating the patient as a whole still remains the first consideration. If this consideration is to be realized, then the training of general accident surgeons is of first importance; once they are properly trained, it would follow that they would delegate to specialists of greater technical experience the treatment of major cerebral and thoracic injuries.

"Stove-in" injuries

Undoubtedly the most serious of all injuries resulting from civilian accidents are the "stove-in" chest with multiple fractures of the thoracic wall and paradoxical respiration, and the "stove-in" pelvis in which great backward displacement of the pubis is associated with considerable upward displacement of the bladder, division of the urethra at the lower level of the prostate and tearing of the anterior wall of the rectum. These "stove-in" injuries call for immediate treatment and are a major exercise in fracture and visceral surgery.

4. ACCIDENT HOSPITALS

The practice of accident surgery is essentially the work of a team of surgeons co-ordinated and in some ways directed by a surgeon-in-charge. The services of the general accident surgical staff must be on a full-time basis, and the essential specialist services on a part-time basis.

Because of the emergency nature of all accident work, the surgical staff must be adequate in numbers. In order that all physical facilities are available for the handling of a large volume of work, an entire hospital is essential, but accident hospitals are perhaps justified only in thickly populated areas—that is, in cities with a population of approximately 200,000. Twenty such hospitals could cover the needs of Great Britain if they worked in close co-operation with numerous "fracture services" elsewhere.

In order that such hospitals should function as economic units they should treat not less than 25,000 patients each year. Their patients would be derived from two sources:

(1) A proportion of the common accidents from the immediate neighbourhood—the type of accident normally treated in a fracture department.

(2) All the serious accidents from the immediate neighbourhood and within 40 miles of the accident hospital. Nowadays such patients are admitted to their nearest hospital by the normal ambulance services but frequently these hospitals have neither the staff nor the facilities for modern treatment. The safe transport of such seriously injured patients to a central hospital can only be effected first by the establishment of a close liaison between hospitals, and next by the provision of a mobile surgical unit with its staff from the central accident hospital. Such a unit has now passed through its trial stage, and on many occasions has proved its value as a life-saving measure.

(1) Organization

In organizing all facilities to meet a large volume of work, for example 30,000 accidents each year, the following requirements have proved necessary.

(i) *Surgical staff.*—Three full surgical teams, each consisting of a surgeon, an assistant surgeon, a registrar, two house-surgeons and a well-qualified anaesthetist.

The Burns Unit

A Burns Unit staff requires two additional surgical registrars and a house-surgeon.

(ii) *Specialist staff.*—The part-time services of cerebral, thoracic, plastic and dental surgeons.

(iii) *Routine pathological and bacteriological services.*—These require a medical director and a small team of technicians.

(iv) *Medical research.*—There is a great deal of research work still needed on the many sides of the accident problem, for example, on the treatment of burns, the healing of wounds, industrial dermatitis, and nutritional disorders in severe injuries. There should be some accident hospitals which provide full facilities and a special staff for medical research.

(2) Building requirements

In building requirements the provision of really adequate facilities in the admission and out-patient departments is the first essential. The admission department particularly must be carefully planned. Through this department all cases of accident, irrespective of their severity, are received by the surgical team on duty, and are followed by that team during their full programme of treatment. Unless the admission department is carefully planned and provided with facilities on a much more generous scale than is usual in hospital buildings, then the regulation of its traffic into smooth, quiet, orderly control is not possible. Unless such control is achieved, the essential careful examination upon which is based all adequate treatment cannot be achieved.

Once patients have passed through an efficient admission department and within that department have received either plenary surgical treatment or surgical first aid, their subsequent treatment can be more smoothly organized [illegible] system ;

[illegible] facilities 90 per cent of the patients can be treated as ambulatory throughout the whole period of treatment. Even the 10 per cent requiring in-patient treatment subsequently become out-patients before their final discharge from treatment. Therefore, the admission and out-patient departments become the key structure of an accident hospital.

Of the 200 beds available for in-patients, 32 should be allocated to the segregated Burns Unit, approximately 10 to the segregated Head Unit and 6 to the Thoracic Unit.

Site of accident hospital

Because of the emergency nature of its work, an accident hospital should be placed in the centre of areas of dense population in a large city. This not only allows the majority of its patients to receive treatment in the shortest possible time, but provides the most convenient facilities for their out-patient re-attendance. However, for long periods of in-patient treatment there are many advantages, both from the viewpoint of the patients' needs and from the hospital economy angle, in the provision of a country branch for the treatment of long-stay patients. One of the most urgent problems in accident treatment is the provision of beds for long-stay fractures in the elderly. Such

patients respond well to modern surgical treatment. The need now is the provision of beds to allow such treatment.

5. REHABILITATION

The rehabilitation of the injured is said to commence with the manner of their ambulance treatment and to end with their resettlement back at work (Griffiths, quoted by Watson-Jones, 1941). This is a catholic conception that has yet to receive a practical interpretation.

The Inter-Departmental Committee placed the whole matter as the responsibility of the treating hospitals. From a theoretical point of view this is an excellent solution, but from a practical standpoint it is economically an impossibility. For economic reasons, rehabilitation and resettlement in their final stages must be provided outside the treating hospitals' buildings.

Accident hospital staffs, after sufficient experience, can become expert in accurate diagnosis, modern surgical treatment and the early and middle stages of reconditioning the patient in preparation for his return to work. They can, if the will is with them, learn something of the nature of the jobs that patients follow in normal health. That knowledge is perhaps essential to a degree in the formation of efficient plans for surgical treatment; for example, it is important to know before removing a torn cartilage from a knee whether the patient is a clerical worker or a miner working at the coal-face. The requirements for full recovery in these two workers differ considerably. The clerical worker must stand and walk and run in order to follow his job and to enjoy his recreational activities. The miner, however, requires much more than this; for him a knee that allows the fullest degree of flexion is essential and he must maintain this position for long periods at work. The requirements of each patient for rehabilitation and final resettlement in work must be known by the treating surgeon, for he alone can make the complete plan for full treatment.

Yet if a hospital is to be given the task of developing muscle and joint movements up to the stage of complete recovery and fitness for work it will be overburdened with patients in their final stages of recovery, and will in consequence have insufficient time to spend on matters upon which it alone is expert.

The final stages of rehabilitation and resettlement are economically and psychologically much better undertaken outside the hospital "atmosphere". There are now two tried methods.

The first is by providing light mechanical engineering jobs *within industry* (Gissane, 1945). When recovery has reached the right stage, the recovering workman is discharged from hospital and is set to work on suitable production machines within industry. Here he undertakes on production machines, slightly modified for his use, the necessary repetition exercises to regain the range, power and speed of movement which are required of his injured extremity in normal employment. The final stage of rehabilitation is placed against productive employment; the worker therefore earns a wage and ceases to become an unproductive member of the community. The hospital, though guiding and to some extent supervising the recovering workman's activities, has handed over its responsibilities to men who have a more detailed knowledge of work requirements, that is, to the shop managers of these

Final stage of rehabilitation

rehabilitation workshops. Having regained the efficiency that is necessary under ordinary working standards the worker is transferred back to his normal work.

The other tried system for heavy workers, such as those in the coal-mining industry, is the establishment of residential rehabilitation centres in the country (Nicoll, 1941), again away from hospital atmosphere, where heavy physical reconditioning can be undertaken as a full-time job in preparation for resettlement at the coal-face.

Both of these final stages of rehabilitation and resettlement involve only a proportion of those patients who suffer from the results of accidents. Hospital rehabilitation can resettle most of the industrial injured, all domestic workers and all children.

In the final stages of rehabilitation and resettlement of disabled industrial workers, the essential aim of the accident hospital should be to work in close co-operation with local industry, and with the resettlement schemes which were initiated by the Ministry of Labour in 1944.

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[References to other titles are given under Injury—Civil and Industrial in the Index Volume.]

INJURY–COMPRESSION

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1. DEFINITION

194.] The term "compression injury" is used as a synonym for "crush syndrome", that is, the sequence of events following ischaemic muscle necrosis after prolonged compression.

2. AETIOLOGY

Air raids

Most reported cases are consequent upon burial underneath debris in air raids, but cases have been recorded following burial in mining accidents and in earthquakes. A similar picture is sometimes seen following rupture or obstruction of a main limb artery in traffic or industrial accidents, due in some instances to ischaemic muscle necrosis, and often called "traumatic anuria".

Traumatic anuria

3. PATHOGENESIS

Muscle rendered totally ischaemic for longer than 3 hours, whether by direct pressure, by a tourniquet or due to arterial lesions, undergoes irreversible necrosis. When the blood supply becomes re-established, the damaged capillary endothelium allows plasma to leak out into the damaged part, which consequently swells. At the same time, the plasma volume becomes decreased and haemoconcentration occurs—as in burns. If this plasma loss is large, the blood-pressure will fall, producing a shock-like state from which recovery will occur on making good the fluid deficit.

The shock-like state

Intracellular substances from the damaged muscle also leak out into the circulation immediately this is re-established. Muscle pigment, potassium, phosphate, creatine and acid are excreted in the urine and may, under certain conditions, produce anuria and irreversible renal failure. It is thought that the mechanism by which renal damage is produced is the same as that following intravascular haemolysis. Experimental work has established that pigment becomes precipitated in the tubules if the urine is highly acid and the flow of urine low. Only in these circumstances can renal failure be produced by muscle substances.

Renal damage

4. PATHOLOGY

In the ...

and opaque, often with a well-marked boundary zone between it and undamaged muscle, corresponding to the edge of the compression marks on the skin. Microscopically, the fibres are necrotic and may become calcified at the boundary zones.

Renal changes

The kidneys in cases in which death occurs from uraemia are swollen and wet. The second convoluted tubules and collecting tubules contain pigmented casts; the first convoluted tubules are dilated and show a mild catarrhal change. The thick segment of Henle's loop shows many ruptures, although the epithelium may show little sign of degeneration. These ruptures allow cast material to be extruded into the interstitial tissue and into veins, as has been demonstrated also in the mismatched transfusion kidney and in acute hydronephrosis. Other pathological changes sometimes found are coincidental (such as fat embolism, central liver necrosis, and pneumonia).

5. CLINICAL PICTURE

Three clinical pictures are seen: the mild case, the severe case without early treatment, and the severe but well-cared-for case. The first and last types show no renal damage. The second type will develop uraemia from which only 33 per cent of the patients recover.

Early recognition

The most important initial point is the history of compression: confirmatory evidence is skin erythema or blistering, muscle paralysis and anaesthesia in the injured area. Later, swelling occurs and the muscle feels doughy. The swelling may become so great as to cut off the peripheral blood supply: gangrene is only averted by incisions into the deep fascia to relieve tension (Belsey, 1942) and by cooling the distal part.

Peripheral gangrene

In severe cases, that is, those with a large amount of muscle involved, "shock" should be anticipated and prevented by isotonic sodium lactate, sodium chloride or plasma infusions, and the haemoglobin level brought down to normal before the blood-pressure begins to fall.

Urine changes

The urine shows the presence of muscle pigment, in solution if the patient has been treated early and adequately by alkali and fluid, or in the form of acid haematin casts or granules if treatment has been inadequate. In the latter cases, the urine is highly acid, reaching *p*H 4 7.

Renal failure

Signs of renal failure are oliguria or anuria, pain in the loins and vomiting; oedema will develop if the uraemic patient is overloaded with saline, tetany if he is poisoned with alkali. Late signs are those of uraemia and of potassium intoxication (irregularity of the pulse and, in the electrocardiogram, increased T-waves and blockage). Some uraemic patients develop a spontaneous diuresis at the end of the first week; if this is large enough, the patient recovers and renal function may return to 100 per cent normal in the course of 6 months.

Recovery

Recovery of the compressed limb depends upon nerve regeneration: the necrotic muscle never, of course, recovers, but its function is often taken over in large part by uninjured portions. Oedema subsides in the third week.

6. DIAGNOSIS

The examination of urine for creatine and benzidine-positive pigment, and the identification of the latter in the Hartridge reversion spectroscope establishes muscle damage.

The chief difficulty in diagnosis occurs in the post-traumatic anuria case (Darmady and his colleagues, 1944), in which the following simulating conditions should be kept in mind:

1. Intravascular haemolysis due to mismatched or overheated transfusion, *Bacillus welchii* infection, soapy abortifacients or burns.
2. Sulphonamide crystal blockage in the ureters or in the tubules of the renal papillae.
3. Renal cortical necrosis (rare).
4. Extrarenal azotaemia due to dehydration or electrolyte loss.

Besides historical and circumstantial evaluation, the following estimations are of value in differential diagnosis: serum bilirubin (or a visual estimate of the colour of the plasma); a qualitative test for urinary chloride (using a silver nitrate in nitric acid solution); examination of urine deposit for crystals, and of urine for sulphonamide, either quantitatively or by the "newspaper test" (one drop of concentrated hydrochloric acid on newspaper soaked in urine and dried gives an orange colour if sulphonamides are present).

7. TREATMENT

As renal failure, once developed, is very resistant to treatment, the first aim is to prevent its development. Prompt restitution of lost plasma and extracellular fluid by transfusion of plasma and solutions of crystalloids such as sodium chloride or lactate is indicated to prevent any fall in renal blood flow. Whether or not transfusion is necessary, a high rate of flow of dilute alkaline urine must be maintained. This is best done by forcing fluid by mouth from the earliest possible moment as a priority measure. Sodium bicarbonate, 1 teaspoonful to a pint of water, should be repeated hourly.

Transfusion

Fluid and alkali administration

Hydration and alkalization can be most rapidly achieved in hospital by the intravenous route, isotonic (1·87 per cent) sodium lactate being used together with glucose saline. If a diuresis is not soon obtained, renal damage is probably already present, and further forcing of alkali and fluid is then dangerous, since alkalosis develops readily in patients with renal failure; if, therefore, an alkaline diuresis is not obtained in the first 12 hours despite reasonable alkali therapy (normal people need about 30 grammes of sodium bicarbonate in 24 hours to retain alkalinity) it should be stopped. In successful cases, alkali should be given at a rate just sufficient to keep the urine alkaline for as long as pigment is being excreted. Such treated cases, of a severity more than sufficient to produce high-grade renal failure if untreated, will put out large quantities of muscle pigment without developing any nitrogen retention. Citrate in quantities larger than 500 millilitres or given rapidly is dangerous.

While most deaths before the fifth day are due to failure to transfuse plasma, or to associated injuries, some have been due to overloading the circulation. Watch should therefore be kept on the neck veins and the lung bases.

Circulatory overload

The use of pressure-bandages to limit plasma loss in the limbs has been advocated (Patey and Robertson, 1942). There seems to be no advantage in this method over that of giving plasma intravenously, and indeed it may possibly be harmful, as in some cases the increase in intramuscular pressure, probably osmotic, due to autolysis, is itself adequate to obliterate traversing

Pressure bandages

blood-vessels against the encircling fascial sheaths. Tight binding would only increase this tendency.

Cooling The injured limb, if at all ischaemic, should be kept cool, rather than, as so often happens, heated. This will decrease the rate of autolysis and also allow living tissue to survive on a smaller margin of blood supply (Allen, 1941)

Immobilization Cooling is ineffective if reactionary hyperaemia has occurred. Immobilization is also needed, as autolytic breakdown and absorption occur more rapidly with exercise of the ischaemic limb.

High carbohydrate intake In the treatment of established renal failure, it is important that nitrogen accumulation should be minimized from the start. This can be achieved by excluding protein from the diet, and giving 2,000 calories per day in the form of butter and sugar, custard, or rice: thus tissue-nitrogen breakdown to supply calories can be eliminated. *Penicillin* Penicillin will control the increased nitrogen metabolism of infection. Sodium sulphate and many other diuretic drugs have been used without signal success. Sometimes spontaneous recovery occurs and then the treatment in use at that time is thought responsible. Theoretically, decapsulation should be successful. In this syndrome, we have not found it so, although some cases of haemoglobin nephrosis have recovered following it. Renal failure of this type is eminently suited for trial of *Artificial kidney* the "artificial kidney" (Kolff and Berk, 1944), as severely affected kidneys have sometimes shown a complete spontaneous recovery. This is a practicable procedure which we have found of use in similar types of acute anuria. *Peritoneal lavage* Peritoneal lavage (Frank, Seligman and Fine, 1946) may also prove to be useful in tiding patients over until their kidneys again begin to excrete.

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INTESTINES

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PART I

INTESTINAL OBSTRUCTION

1. DEFINITION

195.] Intestinal obstruction is the term applied to a failure in the downward passage of the intestinal contents, either from a mechanical occlusion of the lumen or from a fault in the propulsive mechanism.

2. CLASSIFICATION

(1) Chronic intestinal obstruction

Chronic obstruction

This is due to partial occlusion of the lumen, sometimes proceeding to an acute complete obstruction, and a tumour or an inflammatory process in the bowel wall is usually responsible for it.

(2) Acute intestinal obstruction

Acute obstruction

This includes mechanical occlusion, strangulation or interference with the vascular supply and disturbance of the neuro-muscular propulsive force. More than one of these forms of obstruction may be present together: above a mechanical obstruction the bowel may gradually lose its capacity for peristalsis, dilating flaccidly as it does in paralytic ileus; similarly, a strangulated segment of gut, though not occluded, fails to transmit peristalsis and the healthy bowel above it dilates as it would above an occlusion.

3. CAUSES

(1) Simple occlusion

Simple occlusion may be due to obturation of the lumen, disease of the bowel wall or pressure upon the bowel from without.

Obturation may be by foreign body (Storck, Rothschild and Ochsner, 1939), faecolith (Benson and Bargen, 1938), gall-stone (Wakefield, Vickers and Walters, 1939), polypoid tumour or mass of parasites (Kirk and Cantin, 1935). Obstructing diseases of the bowel wall include atresia, tuberculosis, regional ileitis and tumour. Pressure may be produced upon the bowel from without by adhesions, bands, enlarged glands or inflammatory or neoplastic masses *within the abdominal cavity.*

(2) Strangulation

In most forms of strangulation—strangulated hernia, volvulus, intussusception, bands or adhesions—not only are the vessels of the loop occluded but the lumen is simultaneously blocked, yet in these circumstances the strangulation, more serious and rapid in its effects than occlusion, takes precedence in danger and importance. The purest form of strangulation is the early

stage of mesenteric vascular occlusion, which may take the form of arterial thrombosis or embolism, or venous thrombosis. In *mesenteric arterial thrombosis*, the intravascular clot arises usually in relation to an area of atheroma or arteriosclerosis, or in a vessel compressed by a tumour mass. In *arterial embolism* the embolus may derive from a fibrillating left auricle, from a valvular vegetation, from the clot of an aneurysm, from a patch of atheroma or, by paradoxical embolism, from a thrombus in a systemic vein.

Mesenteric vascular occlusion

Mesenteric venous thrombosis may occur as an element in a general portal thrombosis due to cirrhosis of the liver, suppurative pylephlebitis, or splenectomy performed for purpura or splenic anaemia, or it may follow invasion of a mesenteric or intestinal vein by tumour.

(3) Neurogenic obstruction

Neurogenic obstruction may be either paralytic or spastic.

(*a*) *Paralytic ileus*

Paralytic, adynamic or inhibition ileus is a loss of peristalsis, progressive atony and distension of the bowel, due to one of four inhibiting mechanisms.

Inhibiting mechanisms

(i) *Reflex form.*—A truly reflex form may occur rarely after childbirth or aseptic operation; in certain renal diseases (Allen and Jamison, 1940; Svien and Mann, 1943); in torsion of a uterine fibroid or ovary, or of the mesentery or spermatic cord; after minor surgical procedures such as catheterization; on the evening of application of a plaster cast to trunk or pelvis; and after certain injuries, particularly fractures of the ribs or spine, contusion of the abdominal organs and retroperitoneal haemorrhage. The precise nervous mechanism in true reflex paralysis of the bowel is not known, for the condition is rare and not all the reported cases are fully documented to exclude other than nervous influences. It is presumed to be an inhibition of intestinal tone by sympathetic reflex paths, though there is no direct evidence of this except the return of peristalsis which spinal anaesthesia induces by blocking the inhibitory splanchnic paths. Reflex post-operative ileus is probably an exaggeration or prolongation of the abolition of peristalsis which follows all abdominal operations (Moir, 1943; Wakim and Mann, 1943) except those performed under spinal anaesthesia (Davis and Hansen, 1945).

Nervous mechanism

(ii) *Anoxic form.*—An anoxic form of ileus may occur if the bowel is congested from prolonged distension or some other cause; this variety may follow the relief of a mechanical obstruction or of a strangulation.

(iii) *Toxic form.*—The direct effect of bacterial toxins upon Auerbach's plexus is blamed for the paralytic ileus which complicates peritonitis. In many cases, however, this variety of ileus seems to be not purely paralytic, but to be initiated or precipitated by the kinking and twisting of bowel by fibrinous adhesions the occlusive effect of which, though slight, cannot be overcome by the defective post-operative peristalsis; the dilating bowel, working against an increasing mechanical disadvantage, has no chance of recovering its contractile effectiveness. The ileus of peritonitis should in fact be regarded as a combination of paralysis and occlusion. A true paralysis of the intramural plexus seems to be responsible for the meteorism or paralytic ileus of typhoid fever.

(iv) *Chemical form.*—Alterations in the chemical state of the blood may either lessen or augment intestinal contractility. Hypoproteinaemia (Barden

and his colleagues, 1938) lessens peristalsis, as also does a fall in the electrolyte concentration, and these chemical effects may be contributory factors in post-operative ileus.

(*b*) *Spastic ileus*

Spastic ileus, a prolonged and unremitting contraction of a segment or segments of the intestine (Murphy, 1896), is essentially a persistent exaggeration in one or more sites of the contractile phase of peristalsis (Zimmerman, 1930).

Mechanical stimuli

It may arise in response to mechanical stimulation of the mucosa by a gall-stone, a fruit stone or other foreign body, or by a mass of parasites, especially ascaris, or from thermal stimulation by ice-water.

Thermal stimuli

It sometimes occurs from venous congestion, in the early stages of a mesenteric vascular occlusion, for example, or after reduction of a volvulus, intussusception or strangulated hernia. A more truly neurogenic form has been described in cases in which the coeliac ganglion is poisoned by lead, or inflamed or compressed by disease in the adjacent pancreas

Reflex varieties

Other neurogenic causes of spastic ileus are contusion of the abdomen, retroperitoneal haematoma, spinal disease and, more rarely, disease of the vegetative centres of the hypothalamus.

Nervous diseases

Post-operative spastic ileus seems to be the effect of excessive manipulation of the bowel, though occasionally a part may be played in its genesis by hypersensitivity to morphine. A particularly misleading form of spastic ileus is that associated with disease elsewhere in the abdomen—spasm of the small intestine sometimes occurs in association with cancer of the colon, or spasm of one loop of small intestine may accompany herniation of another; spastic ileus may occur in combination with appendicitis, haemorrhage in an ovarian cyst, renal colic or adrenal haemorrhage.

Relation to paralytic ileus

Spastic ileus sometimes proceeds imperceptibly to the paralytic form.

4. MORBID ANATOMY

(1) Chronic obstruction

In chronic obstruction due, for example, to hypertrophic ileo-caecal tuberculosis or to cancer of the colon, the bowel above the obstruction responds to the increased tension within it by a combination of passive dilatation and active hypertrophy.

Passive dilatation and active hypertrophy

The mucosa finally becomes the seat of a catarrhal inflammation, or even of ulceration; perforation of a stercoral ulcer may occur in the dilated, hypertrophied colon above a carcinoma even before occlusion is complete.

(2) Acute obstruction

In acute occlusion, the bowel below the obstruction is emptied by absorption of its fluid content and sometimes by one evacuation. It collapses, and with the loss of stimulating content its peristalsis ultimately ceases; the distal bowel is usually empty and quiet. The bowel above an occlusion dilates with fluid and with gas. The intestinal contents, unable to pass the obstruction, accumulate above it, and their volume is progressively augmented by the continued addition of biliary, pancreatic and gastro-intestinal juices. Their colour becomes darker, changing to brown and ultimately to black due to altered blood which oozes from the congested mucosa. The gas derives from bacterial fermentation (McIver, 1934), from swallowed air (Wangensteen and

Rea, 1939) and from the high gaseous tension in the anoxic blood of congested mucosal vessels. Nitrogen, duffusing from the blood, gradually replaces the oxygen and carbon dioxide, which pass into the blood stream.

Raised intra-intestinal pressure

As the volume of fluid and gas increases, the intra-intestinal pressure rises, and at the peak of peristalsis may reach 75 centimetres of water, soon exceeding the venous pressure in the bowel wall (Elman and Aird, 1935). When the basic or sustained pressure within the lumen rises to 10 centimetres of water, a pressure which is commonly found in the obstructed bowel of man (Sperling, Paine and Wangensteen, 1935), the mucosa becomes congested and oedematous (Sperling and Wangensteen, 1935), absorption comes to a standstill, and still more fluid is poured into the lumen, increasing the intra-intestinal pressure still further. Usually, death occurs before gangrene is established in the bowel wall, but sometimes ulcers occur, or patches of devitalization, on the antimesenteric border at a level where the pressure is particularly high and the wall particularly thin, for example, in the caecum, above a colonic obstruction.

Widespread degenerative changes

After death, degenerative changes may be found in liver, spleen, kidney and adrenal.

(3) Closed-loop obstruction

A special form of occlusion occurs when a loop of bowel is isolated between two obstructions as a closed loop—a situation which arises also if a blind end of bowel, the appendix or some other diverticulum, is obstructed in some part of its length. If the content of a closed loop is sterile, as it sometimes is in appendix or gall-bladder, a mucocele results, but if the content is infected, tension rises very rapidly within the loop, particularly if it is a short one, and devitalization, gangrene and rupture follow quickly. The colon, isolated between a complete obstruction and the competent ileo-caecal valve, behaves as a closed loop; the pressure may rise to a high level, sometimes 70 centimetres of water, and terminal perforation, usually of the caecum, is not uncommon.

Mucocele

Devitalization, gangrene and rupture

Terminal perforation

(4) Strangulation

In strangulation, congestion and anoxia of the strangulated bowel loop are rapidly progressive, the vitality of the bowel is threatened early, and perforation may lead to peritonitis even before the associated obstruction has exercised its general effects. The intestinal infarct is nearly always dark when seen at operation, for in most strangulations the veins are obstructed first; after arterial occlusion there may be a transient pallor, this being recorded in 7 of 359 cases collected by Trotter in 1914, but the dilated capillaries of the strangulated loop fill quickly with blood from the collateral circulation and, by back-flow, from the valveless portal tree.

(5) Paralytic ileus

Paralytic ileus presents at operation or necropsy a gross dilatation of the small intestine, and sometimes of the large intestine, with little or no mechanical cause. When peritonitis has been responsible for its onset, however, evidence of peritonitis is usually still present at the time of death. The lower coils of the ileum, and sometimes the pelvic colon too (the *ileus duplex* of Sampson Handley), are often bathed in a pelvic pool of pus, inflamed, oedematous and

distorted and kinked by slender fibrinous adhesions the mildly adhesive effect of which the inadequately contracting bowel has been unable to overcome.

(6) Spastic ileus

If a patient suffering from spastic ileus undergoes operation the spasm usually relaxes under anaesthesia, and distended bowel is seen to merge imperceptibly into collapse, without any detectable cause of obstruction. Sometimes the spasm persists at operation, particularly if a spinal anaesthetic is used, and a few inches, or even a considerable length, of ileum or of colon, or of several segments, may then be found contracted to the limit, white, bloodless, and so firm that the affected bowel may be raised by one end and held horizontally like a lead pencil (Freeman, 1918). The contraction ceases abruptly at either end and the remainder of the intestine is normal unless the spasm persists for so long that the proximal bowel is forced to dilate.

5. MORBID PHYSIOLOGY AND GENERAL EFFECTS

(1) Water and salt loss

In high occlusion

Though perforation and peritonitis are dangerous in colonic obstructions and in strangulations, the most serious effects of intestinal occlusion and of the neurogenic varieties of obstruction are dependent upon the loss to the organism of water and salts (Haden and Orr, 1923; Armour and his colleagues, 1931). These cannot pass beyond the obstruction to be absorbed, as they normally are, at a lower level, but accumulate above the obstruction in distended bowel the congested wall of which is incapable of absorbing them. The lower the obstruction and the greater the length of absorbing surface and distensible bowel above it, the later the onset of serious water and salt loss; the most serious and rapid loss occurs in obstructions just below the ampulla of Vater, the *lethal line* of Maury (1909), for in obstruction at this level all the salivary, gastric and pancreatic juices and the bile are lost. This fluid sometimes amounts to 8 litres daily in a man weighing 70 kilograms (Adolph, 1933). Obstructions at lower levels produce a depletion of water and salt which, though still important, is slower and less serious. In strangulation, other and more rapidly lethal factors are in operation, and the patient may die before water and salt loss is sufficient to be serious.

Lethal line

In low occlusion and strangulation

(2) Compensatory mechanisms

Compensation for water loss

The water and salt lost in high occlusions passes to the bowel in the first place from the plasma. The consequent tendency of the plasma volume to fall is at once corrected by a reduction in renal secretion and by the passage of water from tissue spaces (interstitial fluid) to blood, attracted by the rising tendency of the plasma osmotic pressure.

Compensation for chloride loss

The chloride loss is made good partly by a retention of chloride from the urine, partly by a passage of chloride from corpuscles to plasma, partly by bicarbonate replacement of the lost chloride and partly, but only partly, by the passage of chloride from tissue spaces to blood. Proportionately less chloride than water thus passes from tissue spaces to blood; the osmotic pressure in the extracellular fluid consequently tends to rise and water is withdrawn from the cells. Thus the loss of chloride is at first mainly from the interstitial fluid whereas the loss of water is from interstitial and intracellular

fluids; the intracellular fluid is materially, the interstitial fluid substantially, reduced in total volume, but the plasma volume remains unaltered and the plasma chlorides only slightly altered, until a late stage of the disease is reached; laboratory methods at present available are poor criteria of the degree of water and salt loss. The only reliable evidence of dehydration in the patient who presents with early intestinal obstruction is clinical (*see below*).

(3) Terminal chemical changes in the blood

Only in the last stages of intestinal obstruction does the blood chemistry alter. The changes in it include: reduction of plasma volume, high haematocrit reading, increase in haemoglobin, red cells and plasma protein, and decrease in chloride (though chloride loss is sometimes obscured even at death by a corresponding decrease in plasma volume, the chlorides remaining at quite a high level).

Blood concentration

Hypochloraemia

When chloride loss occurs, the threatened fall in electrolyte concentration is compensated for by bicarbonate retention; the bicarbonate content of the blood rises, as does the carbon dioxide combining power, and the hydrogen ion concentration finally falls.

Alkalaemia

Meanwhile, and often before other chemical signs of dehydration are available, the nitrogen metabolism suffers a remarkable change; the blood urea and the non-protein nitrogen rise. The elevation of these nitrogenous elements seems to depend not only directly upon dehydration but also upon two additional factors: (1) with dehydration the urinary output falls and the secretion of a concentrated urine from a concentrated blood affects the efficiency of the kidney; the rise in blood urea is a sign, in some cases, of renal damage, which may be proved by the presence of albumin, red cells and casts in the urine: (2) cellular dehydration may be accompanied by increased protein catabolism and a consequent liberation of urea.

Azotaemia

These effects of dehydration have been discussed at some length, since it is important to appreciate that the blood changes are usually late, sometimes contradictory and often misleading; it is invariably dangerous to use them as a rigid index of the state of hydration of the patient, or as a basis for calculation of his water and salt needs.

(4) Depletion of blood volume

Factors other than fluid and salt loss play a part in the physiological disturbance which accompanies certain forms of intestinal obstruction. A substantial fraction of the blood volume may be held up in the congested vessels of distended bowel. In extensive strangulations, the blood volume may be reduced by as much as 50 per cent (Aird, 1935b, 1937 and 1938), the lost blood lying stagnant in the vessels of the strangulated loop, and being poured into the lumen of the strangulated bowel and into the peritoneal cavity.

Stagnant blood

(5) Toxic factors in strangulation

The absorption of toxins from distended bowel loops has not been shown to be responsible for any part of the disturbed physiology of obstructed patients, but toxic absorption *is* important in strangulation (Holt, 1934). In strangulation, the fluid poured from the congested bowel into the peritoneal cavity has a toxic vasodepressor effect (Aird, 1936a and b ; Knight and Slome, 1936), not lethal in itself, but perhaps serious in an acutely anaemic

Toxic vasodepressor effect

subject (Maycock, 1938). The peritoneal transudate has been shown to owe its toxicity to its protein content, and to the vasodepressor effect of the histamine (Aird and Henderson, 1937) and choline (Maycock, 1938) which it contains. The toxic effect of strangulation resembles that of moist gangrene; *intestinal strangulation is in fact an intra-abdominal form of wet gangrene;* its toxicity is due to bacterial invasion of the infarcted bowel; there is no toxaemia in strangulation of the sterile gut of a new-born animal.

Form of wet gangrene

6. CLINICAL PICTURE

Pain

The first symptom of acute intestinal obstruction is pain; pain is absent only in the paralytic variety. The pain is colicky and median though in strangulation it may be referred to the position of the strangulated loop if the serous coat becomes inflamed.

Constipation

In complete obstruction there is complete constipation, and neither flatus nor faeces is passed except just after the onset, when there may be one bowel movement. In obstruction of the rectum or lower pelvic colon there may be tenesmus, and in intussusception, mesenteric vascular occlusion or volvulus a blood-stained stool may be passed. If an enema is given it is returned clear or slightly coloured, with perhaps one or two scybala but no flatus.

Melaena

Vomiting

Vomiting is early in high obstruction, later in low obstruction, and in colonic obstruction it may be delayed for some days. First, recently digested food is



FIG. 40.—Peripheral distension of colonic obstruction immediately after relief by caecostomy.



FIG. 41.—Distension of flanks in colonic obstruction.

vomited, then clear mucoid fluid, then bile, and finally the black and malodorous fluid which, formerly called faecal, owes its colour and smell to altered blood.

Signs of dehydration

On physical examination the patient may already, when first seen, present the dry tongue, glazed sunken eyes and thin dry skin typical of dehydration. The urine is concentrated and small in amount with little if any chloride. These signs, together with thirst, distension and a history of vomiting, are

arer evidence of the presence of dehydration than any chemical tests at esent available.

On examination of the abdomen, distension may be observed but is con- lerable only in low obstructions. In ileal obstruction the distension is ntral and the obstructed coils may lie one above the other as a ladder ttern; in colonic obstruction the distension is mainly peripheral at first d the central distension of the small intestine is superimposed upon the ripheral pattern only in the later stages of the disease (Figs. 40 and 41). sible peristalsis may be elicited, but its presence is in itself no indication of istruction, for a thin abdominal wall may allow normal peristalsis to be en, nor is its absence proof of the absence of obstruction.

Distension

Visible peristalsis

The hernial orifices should always be inspected, for the commonest cause of istruction is external hernia and the patient may not know that he has a pture.

Hernial orifices

Palpation of the abdomen may reveal a tumour mass, inflammatory tender- ss, the rebound tenderness of peritonitis or strangulation, the sausage- aped swelling of an intussusception, or the tense, massive, tympanitic lloon of a volvulus.

Palpable mass

Physical examination is complete only after digital examination of the ctum has been performed. This may demonstrate a rectal cancer or a alignant or inflammatory mass in the pouch of Douglas. The examining nger may be blood stained in intussusception or mesenteric vascular occlu- on; indeed in a low intussusception the apex may be palpable or visible from e anus.

Rectal examination

7. SPECIAL AIDS TO DIAGNOSIS

1) Radiography

A "scout" or straight skiagram of the abdomen may indicate not only the resence of obstruction but also its level and sometimes its nature. Gas- lated loops may be recognizable as small or large intestine, and so indicate e level of obstruction; the film may demonstrate the shadow of a gall-stone bstructing the ileum, or the presence of an opaque foreign body.

2) Diagnostic enema

A diagnostic enema of half a pint of warm saline or soap and water may be dministered. If this is returned only slightly coloured and unaccompanied y flatus the obstruction may be assumed to be complete, though a flatus eturn may be obtained if the obstruction is of very recent origin. If the enema s returned blood stained a strangulation should be suspected—intussuscep- ion, volvulus or infarction (Cokkinis, 1926) A repeat film of the abdomen fter the enema will demonstrate how effectively the intestinal gas has been vacuated (*see section below*).

8. DIFFERENTIAL DIAGNOSIS

1) Presence or absence of obstruction

Obstruction may be presumed to be complete if a diagnostic enema is re- urned without faeces or flatus, but an absolute opinion cannot always be based upon this information alone. The information obtained by enema should be supplemented by that obtained from skiagrams. Briefly it may

Completeness of obstruction

be said that if small-intestine loops are distended by gas while the colon is free of gas or emptied successfully of gas by enema, a *complete small-intestine obstruction* is present. If the colon or any part of it remains gas-distended after diagnostic enema, a colonic obstruction is present. In incomplete obstructions gas may be seen in bowel loops both above and below the level of obstruction. In a partial obstruction of the ileum, for example, not only are the loops of the small intestine distended with gas, but gas is radiologically demonstrable in the colon too; the colonic gas may be evacuated by enema but is shortly replaced again from above.

(2) Distinction from other acute abdominal diseases

Inflammations of hollow intraperitoneal organs, cholecystitis and perforative peritonitis, can usually be excluded by the relative absence of muscular rigidity; in the later stages of perforative abdominal diseases, of course, peritonitis and ileus are combined. Extreme constipation may closely simulate acute obstruction, particularly if impacted faeces or resinous foods (persimmons or medlars) become tightly lodged in the colon; in such circumstances a diagnostic enema is rarely inefficacious, and sufficient flatus is passed to prevent serious distension.

(3) Distinction between occlusion, strangulation and neuro-muscular disorder

The needs of treatment demand that these conditions be clearly distinguished, for strangulation demands immediate operation, occlusion requires vigorous and sometimes prolonged preparation for operation, and neurogenic obstruction is best managed without any operation at all.

Occlusion distinguished from strangulation

In both occlusion and strangulation there is pain, but in strangulation the pain is more severe and the spasms are more frequent and regular, returning several times a minute with increasing rather than lessening intensity. The chief distinction between strangulation and occlusion, however, is the presence or absence of rebound tenderness Sometimes in occlusion, as always in strangulation, there is tenderness when the hand is depressed into the abdomen over an obstructed loop, but in occlusion elevation again of the examining hand is as painful as its depression. The rebound tenderness of strangulation depends upon inflammation of the serous coat of the strangulated loop; it cannot be elicited readily over an intussusception, for the intussusceptum is protected from the palpating hand by the sheath.

Ileus

Paralytic or adynamic obstruction is characterized by an entire absence of colic and by dead silence of the abdomen on auscultation. Spastic ileus may be suspected if the signs of obstruction are associated with a scaphoid rather than a distended abdomen; sometimes the pulse rate is strikingly slow, being only 60, 50 or even 40 per minute.

(4) Level of obstruction

High obstruction

In high obstruction vomiting is early and distension late, and the straight skiagram after diagnostic enema may show gaseous distension only of the uppermost jejunal loops or of the duodenum alone.

Low obstruction

In low small-intestine obstruction vomiting may be relatively early. Distension is considerable and central in position, often accompanied by ladder pattern and multi-directional peristalsis. The straight skiagram shows

gas-distended loops of small intestine with or without multiple fluid levels. The colon may or may not be emptied of gas at the time of the first skiagram, but it will be in a second film exposed after a diagnostic enema.

Colonic obstruction In colonic obstruction vomiting is late, sometimes being deferred for many days, and distension is peripheral. An exception to this rule is volvulus of the pelvic colon, for the twisted and distended pelvic loop often assumes a central situation. The level of colonic obstruction can usually be determined accurately by a "scout" film. Low rectal obstruction should be detected by the finger.

(5) Nature of obstructing lesion

The cause of obstruction can often be precisely determined before operation

Age as a guide At birth, atresia, and a few days or weeks later, volvulus neonatorum, are the commonest causes of obstruction. In the second and third months incarcerated hernia is most likely to be responsible, and from the third to the twelfth month intussusception. In later childhood plastic tuberculous peritonitis is not infrequently to blame. In early adult life post-appendicectomy adhesions are a common cause of obstruction, though tuberculosis of bowel or peritoneum may occur in this period too, and strangulated hernia begins to be important. Men in the thirties and forties suffer particularly from strangulated inguinal hernia, women in the forties or later from strangulated femoral or umbilical hernia or gall-stone ileus, though this last disease is less common now than formerly. In the older age periods cancer of the colon is the commonest cause of obstruction, with strangulated hernia only second in importance.

(*a*) *Strangulated hernia*

The cause is obvious if a tender external hernia is found in the course of examination, and inspection of the hernial orifices, common and rarer, should never be omitted. Strangulated internal hernia is more difficult to detect, but the tense strangulated loops give a typically localized, tender, tympanitic prominence. The presence of an operation scar suggests that an adhesion or band is responsible.

(*b*) *Cancer*

Colonic obstruction should always be regarded as malignant unless some other cause can be proved; the obstructing tumour is sometimes palpable as a hard, irregular and perhaps tender mass. Most malignant obstructions are preceded by some weeks or months of increasing partial obstruction, with constipation or diarrhoea or both and often with dyspepsia, before occlusion finally becomes complete.

(*c*) *Gall-stone obturation*

If the patient is a stout, middle-aged woman with a dyspeptic history suggestive of chronic cholecystitis and a recent attack of upper abdominal pain suggestive of acute cholecystitis, it may be suspected that a gall-stone has impacted in the lower ileum, having passed from gall-bladder to duodenum by fistula during the recent acute cholecystitis. A straight skiagram sometimes shows the shadow of the impacted stone, usually in the right lower quadrant, with gas-distended loops related to it (Lowman and Wissing, 1939)

Sometimes the obstructive symptoms occur in a series of episodes which follow at short intervals a severe attack of cholecystitis, the stone passing into the duodenum and then proceeding down the intestine, being temporarily held up at level after level before firmly and finally impacting in the lower ileum.

(*d*) *Mesenteric vascular occlusion*

When obstruction is accompanied by rapid collapse, increase in pulse rate, fall in blood-pressure and signs of internal haemorrhage, mesenteric infarction should be suspected. Suspicion hardens to certainty if the patient suffers from auricular fibrillation, mitral stenosis, endocarditis, atheroma or aneurysm. Sometimes proof is obtained by the return of blood in a diagnostic enema (Cokkinis, 1926).

(*e*) *Intussusception*

History

Intussusception is easy of diagnosis if the patient is a lusty child a few months old, with a history of sudden pain, updrawing of the legs on the abdomen and the passage of blood, but even in an adult, when the intussusception is usually secondary to simple tumour, the sausage-shaped swelling is hardly mistakable.

(*f*) *Volvulus*

Volvulus of the pelvic colon may be suspected if a man in middle life presents the signs of strangulation with or without the passage of blood, but accompanied by palpable distension of the sigmoid colon.

9. TREATMENT

(1) General management

There are three measures available for the treatment of intestinal obstruction: gastro-duodenal suction, intravenous fluid administration and operative correction. Given early diagnosis, success in treatment depends upon the prompt commencement, the efficient management and the adequate continuation of the first two of these, and the choice of a proper moment for operative intervention.

Occlusion of the small intestine

When a confident diagnosis of occlusion of the small intestine has been made, gastro-duodenal suction drainage is established, the patient's requirements of water and salt are estimated and intravenous fluids are given in suitable amounts and proportions to restore the fluid balance in 24 or 48 hours. An input and output chart is kept, and the progress of distension is followed by repeated inspection and by daily x-ray examination, which should show a progressive decrease in the area of gas shadows. Provided diagnosis is accurate, strangulation confidently excluded, water and salt balance preserved, distension shown clinically and radiologically to be diminishing daily, and pain constantly absent, this conservative regimen may be continued indefinitely. In some forms of adhesive obstruction it will be found possible, after distension is relieved, to clamp off the tube for 3 or 4 hours at a time and to allow fluids by mouth; if there is no discomfort, no distension and no vomiting, the tube may be removed, to be replaced if discomfort, clinical distension, radiological gas accumulations or vomiting return. In most small-intestine occlusions, however, operation should be undertaken when distension is relieved and lost salt and water replaced.

If pain persists for more than 1 hour after the establishment of effective gastro-duodenal suction it should be assumed that the clinical diagnosis of simple occlusion is faulty and that strangulation is present or threatened; operation should then be performed at once. Operation is indicated also if pain returns during treatment, if distension fails to decrease in spite of effective suction and if skiagrams taken on successive days show identical or increasing gas shadows in the small intestine.

Colonic obstruction

In colonic obstructions there is less dehydration, and the risk of perforation in a greatly distended colon makes early operation advisable.

Strangulation

If strangulation is suspected from the symptoms and signs, and particularly if rebound tenderness is present or if pain persists for more than an hour after the establishment of effective gastro-duodenal drainage, operation should be undertaken forthwith, and delayed only for the passage of a duodenal tube (if that has not already been done) and the setting up of an intravenous drip not only of saline but of blood, for extensive strangulation seriously depletes the blood volume (Scott and Wangensteen, 1932; Aird, 1935b, 1937 and 1938).

Mesenteric vascular occlusion

The only form of strangulation in the treatment of which operation is sometimes omitted is mesenteric vascular occlusion. Operative resection is still attended by a high mortality; Whittaker and Pemberton (1938) lost 16 patients out of 19 and their experience is happier than most. On the other hand, spontaneous recovery is by no means uncommon. There is a strong argument for the treatment of mesenteric vascular occlusion by full heparinization (Murray, 1940), gastro-duodenal suction and intravenous transfusion of saline, plasma and blood. Even if resection is performed, post-operative heparinization restricts the spread of thrombosis.

Paralytic ileus

Paralytic ileus is treated also in a conservative way, by the same measures as are used for its prevention in patients suffering from peritonitis. Suction drainage is begun and an intravenous drip set up. No active measures are taken to induce a return of normal bowel movement. With this treatment vomiting stops, distension disappears and dehydration improves. As in occlusion, the progress of gaseous distension is estimated daily, clinically and radiologically. When the abdomen is again flat, the suction tube is clamped for 3 or 4 hours at a time and liberal drinks are permitted; if distension or vomiting returns, suction is re-established. If there is no return of distension or vomiting, the tube may be withdrawn and left out overnight. In most cases, flatus is passed spontaneously or after a small enema. Sometimes distension or vomiting returns after removal of the tube, which should then be reinserted.

Jejuno-transverse colostomy for persistent post-operative ileus

After a fortnight of conservative treatment, in the ileus of peritonitis, if flatus has not yet been passed and distension returns when the suction tube is clamped or removed, and particularly if peristalsis is audible, it may be assumed that a persistent degree of mechanical obstruction is present. This can be relieved easily and safely, and invalidism shortened, by the operation of jejuno-transverse colostomy, for the obstruction seems usually to be in the jejuno-ileum. The bowel having been decompressed by suction, and water and chloride balance well adjusted, the abdomen is opened and an anastomosis is performed between a presenting loop of distended jejunum and the transverse colon, which is always at this time found to be collapsed. This operation

should not be too long delayed; it is followed usually by an interval of several days without bowel action, but in most cases the suction tube may be removed on the morning after operation without recurrence of distension. The patient's general condition, affected—in a fashion more easily recognized than described—by several days of purely intravenous alimentation, improves at once.

Spastic ileus

Spastic ileus, whether detected at operation or before it, is best treated by atropine and intravenous pethidine, the latter in doses of 100 milligrams hourly until the spasm is relieved. Here too, suction drainage and intravenous fluids should be afforded.

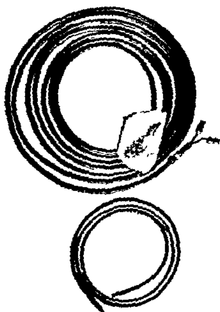


FIG. 42.—Ryle and Miller-Abbott tubes for gastro-intestinal suction

(2) Gastro-duodenal suction

Gradual deflation

Gastro-duodenal suction, as Westermann first showed in 1910, effectively relieves intestinal distension. Gradual pre-operative deflation has several advantages—it facilitates operation, reduces operative shock, lessens congestion and oedema in the bowel wall, and cancels the dangers which have been shown to attend the sudden operative deflation of distended bowel (Elman and Aird, 1935). A Miller-Abbott (Fig. 42) or Wangensteen tube (Abbott, 1941; Wangensteen, 1942) is passed, and suction by pump or siphonage is maintained before and after operation, or without operation if no operation is required, until its cessation is no longer followed by distension, clinical or radiological, or by vomiting. Gastro-duodenal suction is always combined with the continuous intravenous administration of fluid; suction removes vast quantities of fluid and salt from the body.

Miller-Abbott and Wangensteen tube

Technique of intubation

The main technical difficulty in intubation is the negotiation of the pylorus. The balloon-ended Miller-Abbott tube passes more easily if tipped with a mercury bag or a 12-inch whip-lash of catgut, or if stiffened by 2 feet of 0·4 millimetre piano-wire let into the tube just proximal to the balloon. When first passed, the tube should not be paid out all at once. The tube is passed into the stomach, which is emptied by a short period of suction. The balloon is then slightly distended and passed to a total distance of 30 inches. No more should be paid out until a skiagram shows the tip to be in the duodenum; the balloon may then be fully distended and the tube passed to its limit. The indwelling tube is well tolerated. Infants decompressed by it sometimes develop otitis media, but this is a common complication in dehydrated babies; Iglauer and Molt (1939) have recorded laryngeal necrosis and oedema from pressure of the tube upon the anterior wall of the hypopharynx, but this complication does not seem to arise in a conscious patient who swallows frequent sips of sterile water.

Fine, Banks and Hermanson (1936) showed that the prolonged inhalation of 95 per cent oxygen may have a decompressive effect by reducing the tension of nitrogen in the plasma and consequently in the bowel, but this decompressive effect is slow.

(3) Maintenance of water and salt balance

The intravenous administration of fluid is begun as soon as any patient suffering from obstruction is seen, and is continued for so long as gastro-duodenal suction is maintained, and until fluid can be assimilated by mouth.

The first day's fluid

As has already been explained, the salt and water needs of a patient suffering from intestinal obstruction are difficult to estimate at the time of first assessment. The biochemist cannot precisely measure the degree of dehydration by methods at present available, and the administration of salt and water must thus be empirical during the first 24 hours after the patient's admission to hospital. It may be assumed that a patient suffering from complete obstruction of the small intestine, and presenting manifest evidence of dehydration, has lost 6 per cent of his body-weight (Coller and Maddock, 1940); a patient weighing 70 kilograms requires 4 2 litres to repay this debt, with in addition some 200 millilitres per hour to replace the intestinal juices withdrawn by suction. Thus, if it is planned to correct dehydration over a period of 24 hours, a total of 8 litres of intravenous fluid is required, or approximately 108 drops per minute.

Not all of this fluid should be given as saline. The salt loss is proportionately less than the water loss, and the chemical process of compensation for chloride loss, should not, and indeed cannot, be rapidly reversed; saline too rapidly given may raise dangerously the filling pressure of the right heart; and the kidneys, tried by dehydration, may not excrete an excess of chloride quickly enough, the interstitial fluid may become hypertonic and oedema may develop, manifestly in the extremities and dangerously in the lungs. If, however, the first and then every third litre is given as normal saline and the remainder as 5 per cent dextrose there is no danger either of oedema or of overloading, for unless the kidneys are the seat of gross antecedent disease they can excrete the water, though not the salt, as rapidly as it can be administered.

Fluid balance of the past day

After the first and every subsequent period of 24 hours the water and salt balance of the past day is closed, and the budget of the succeeding day is planned (Fig. 43). The fluid output (Coller and Maddock, 1940; Bingham, 1938; Abbott, 1946) is the sum of (1) the total volume of urine passed, (2) the volume of gastro-intestinal juice withdrawn, (3) 500 millilitres in respiratory loss, (4) 500 millilitres in perspiration. Against this is balanced the intake—the amount given by intravenous drip plus the fluid taken by mouth. The resultant credit or debit is carried forward to the next day.

Fluid needs of the coming day

It may be assumed that the gastro-intestinal juice removed by suction will be the same during the new day as in the day just past. To this is added 1,500 millilitres for urine, 500 millilitres for respiratory loss and 500 millilitres for perspiration, plus a further 500 millilitres if the patient is febrile. This amount, plus or minus the debit or credit from the previous day, is the volume of fluid required during the subsequent 24 hours. The number of litres required,

HAMMERSMITH HOSPITAL: BRITISH POSTGRADUATE MEDICAL SCHOOL.
DEPARTMENT OF SURGERY.

FLUID BALANCE CHART.

NAME: WHITE

DATE: 4.3.47 – 5.3.47

TODAY'S SALT NEED

Yesterday's balance ± 5 gm
Basal need . . . 5 gm
Salt loss in juices (0 5 per cent of vol.) . . 6 gm
If pyrexial, add 5 gm. . gm.

Total salt needs today . 16 gm —expressed as normal (0 9 per cent) saline = 2 litres
Remainder as 5 per cent dextrose = 3 litres

TODAY'S FLUID NEED

Yesterday's balance ± 2000 ml
or on 1st day, 4200 ml : — ml.
Juices lost, as yesterday, 1200 ml.
Urine, lungs, skin: 2500 ml
Plus 500 if pyrexial ml.

Total fluid needed: 5700 ml.
By mouth, at ml. per hour: 720 ml

TOTAL INTRAVENOUS FLUID: 5 litres
Drip rate per min. (litres × 12): 60 drops

INTAKE					*OUTPUT*				
		INTRAVENOUS					JUICES		
Time	ORAL	*Saline*	*Dextrose*	*Plasma or Blood*	Time	URINE	*Suction*	*Vomit*	*Enter-ostomy*
hrs.	ml	ml	ml	ml	hrs.	ml	ml.	ml.	ml.
09.00	60	1000							
10.00	60								
11.00	60								
12.00	60				12.45	300			
13.00	60	1000							
14.00	60								
15.00	60								
16.00	60								
17.00	60								
18.00	30		1000		18.15	300			
19.00	30								
20.00	30								
21.00	30						600		
23.00			1000						
05.00			1000		06 00	350			
07.00	30								
08.00	30						1200		
Total	720	2000	3000		Total	950	1800		

TOTAL INTAKE 5720 *ml* *URINE AND JUICES* OUTPUT (U J) 2750 ml.

BALANCE AT END OF DAY

SALT Yesterday's balance . ± 5 gm.
Basal . . . . . 5 gm.
Juices (litres × 5) . 9 gm
If pyrexial add 5 . gm.

Today's total salt usage 19 gm
Subtract total intake (litres × 9) . 18 gm

Carry forward balance . ± 1 gm

WATER. *Yesterday's balance* ± 2000 ml.
Skin, lungs today . 1000 ml
Add 500 if pyrexial . — ml.
Today's output (U.J) . 2750 ml.

Today's total usage . 5750 ml.
Subtract total intake . . . 5720 ml.

Carry forward balance ± 30 ml.

FIG. 43.—*Specimen daily chart of fluid balance during treatment for intestinal obstruction* Note previous day's balance at top to decide daily intake, balance below of day just finished to carry over to next sheet

multiplied by twelve, gives approximately the number of drops per minute at which the drip should run.

Salt needs

The salt needs of the patient are satisfied by 5 grammes of sodium chloride in the day, plus a further 5 grammes for every litre of gastro-intestinal juice withdrawn; only sufficient normal saline is given to cover these amounts of salts, and the balance of fluid required is given as 5 per cent dextrose.

Fluid by mouth

So long as suction is continued, water may be taken by mouth, but in rather restricted quantities—not more than 4 ounces in the day. Most of the water swallowed is removed by the tube, and constant lavage of stomach and duodenum may wash out salt in uncontrolled amounts and upset the salt balance.

(4) Details of operation

(a) Anaesthesia

For the exploration of an intestinal obstruction spinal anaesthesia gives complete relaxation, but the immediate encouragement which it gives to peristalsis is of less certain value. The rapid deflation of congested bowel may give an alarming fall of blood-pressure which is perhaps responsible for some operative and early post-operative deaths. The two disadvantages of inhalation anaesthesia, inadequate relaxation and copious vomiting, are less important now that curare affords relaxation, and pre-operative suction lessens the risk of vomiting.

(b) Incision

The incision should afford ready access to the expected site of obstruction, in most cases a right paramedian incision is suitable.

Operative diagnosis

Once the abdomen is opened the hand is passed first to the suspected site of obstruction; the actual cause may then at once be obvious. If the site is not known, the hand is passed first to the caecum. If the caecum is collapsed, the lowest loop of ileum is withdrawn, and collapsed bowel is followed upwards, loop by loop, to the obstruction. If the caecum is distended the pelvic colon, and then the transverse colon, is palpated, the condition of these indicates the level of colonic occlusion.

(c) Relief of obstruction

Simple strictures

Tumours

Foreign body

Once the obstruction is found, appropriate measures are taken for its relief. Bands or adhesions are divided, care being taken to deal with all multiple constrictions. Simple strictures are short-circuited by lateral anastomosis rather than excised. Tumours of the small intestine are immediately excised only if the occlusion or intussusception which they have produced is recent, and the distension of slight degree; otherwise anastomosis should be performed and resection postponed. An obturating gall-stone, faecolith or foreign body is best crushed between the fingers—if that can be done without bruising of the bowel wall—or passed onwards or backwards for removal through a portion of bowel distal (preferably) or proximal to the often overstretched and sometimes devitalized site of impaction The treatment of intussusception, volvulus and hernia is described elsewhere.

(d) Enterostomy

Irremovable obstruction of the small intestine is best treated by lateral anastomosis. The fluid loss of an enterostomy is almost as serious as its discomfort is trying If, however, the patient's condition is serious and

distension unrelieved by [illegible]
a life-saving measure. A [illegible]
remainder of the intestin[illegible] applied to occlude doubly the presenting loop which is then emptied by aspiration through a wide-bore needle. A purse-string suture of catgut is then inserted round the site of needle-puncture in a circle 2 centimetres in diameter, and a stab incision is made to enlarge the puncture wound. Into this is passed a red rubber catheter (size No. 14 French) disto-proximally for 5 centimetres. The purse-string is drawn tight, tied, and tied again round the catheter. The catheter is then laid along the bowel and buried by a continuous catgut suture for a distance of 5 centimetres, and the end of the catheter, threaded through omentum, is drawn out through a stab wound in the abdominal wall so that the drained loop fits snugly against parietal peritoneum. The catheter is closed by a clip, the clamps are removed, the abdominal wound is closed and dressed and the catheter is then allowed to drain.

(e) *Colostomy or caecostomy?*

Colonic obstruction

Irremovable obstruction of the right colon is usually best treated by ileo-transverse colostomy, that of the left colon by transverse colostomy and that of the lower pelvic colon or rectum by pelvic colostomy. Yet here, as in the small intestine, if the patient's condition is serious and the colon grossly distended, external drainage may be performed by the quickest possible means—in this case caecostomy. The distended caecum is withdrawn through a separate grid-iron incision in the right iliac fossa, and emptied by the gentle pressure of one hand while the other hand applies a curved clamp across the caecal base. A purse-string suture is inserted, a stab incision is made in its centre and a small glass colostomy tube is inserted. A second purse-string is inserted and drawn tight at a higher level on the tube than the first, and the caecum is anchored by two sero-muscular stitches to the parietal peritoneum of the grid-iron incision.

Criteria of viability

In all strangulations the operator must, after relief of the obstruction, decide whether the affected bowel is viable or not. If viable, the bowel should be returned to the abdomen. Small intestine, if not viable, should be resected; non-viable large bowel should always be exteriorized rather than resected immediately.

The bowel may be regarded as viable if its sheen is still present, if its colour is pale or pink, if it transmits peristalsis and if the vessels in its mesentery pulsate. Bowel is non-viable if its sheen is lost; if it is purple, dark green, persistently dark blue or black in colour; if it emits a detectable odour; if it fails to conduct peristalsis, or if the vessels in its mesentery are thrombosed.

If the signs of viability are at first absent the bowel should be wrapped for a few minutes in a warm towel while the anaesthetist floods the lungs with oxygen to encourage the return of its colour. To excite peristalsis the proximal bowel is gently flicked with the finger. In examining a suspicious loop special attention should be paid to its ends where constriction rings are of peculiar danger. In general, doubtful bowel may be returned to the abdomen if the patient's condition is poor, for even the most apparently gangrenous loop, if limited in length, may recover within the abdomen, though with the risk of stricture formation later (Tanner and Bratton, 1943).

10. PROGNOSIS

The longest series of cases of intestinal obstruction are those collected by Souttar (1925) and Vick (1932). The general fatality rate in Vick's composite series of nearly 7,000 cases collected from the 1925–30 records of 21 British hospitals was 26·2 per cent. The percentage fatality rate in each group was as follows: enterolith, nil; inguinal hernia, 11·5; femoral hernia, 15·9; primary intussusception, 17·6; all hernia, 17·8; foreign bodies, 20; faecal masses, 23·5; secondary intussusception, 26 7; adhesions, 32·9; inflammatory stricture, 33 3; internal strangulation, 34·3; external hernia other than femoral, inguinal or umbilical, 37; umbilical hernia, 39·9; sigmoid volvulus, 41; tumour compression, 41·4; cancer, 42 6; volvulus of caecum, 45·7; congenital stricture, 57·5; enteric volvulus, 62·4; gall-stone obstruction, 70 2; embolism and thrombosis, 91 8.

The great diversity of fatality rate in these 21 different varieties of obstruction shows how important it is that any series of cases used for statistical purposes should be long. The series quoted above was collected before the introduction of saline therapy and suction drainage; it is likely that a comparable series compiled 10 years later would show a decrease of perhaps 10 per cent in the total mortality rate. Short series of one or two hundred cases published in the last few years suggest that this is so. In some quarters the introduction of suction drainage has resulted in a fall in the fatality rate of occlusion but a rise in that of strangulation, operation in the latter condition sometimes being dangerously delayed by failure to distinguish it from occlusion.

It will be remarked that though enterolith obstruction resembles gall-stone ileus in its mechanism, the fatality rate is nil in the former and 70 per cent in the latter. The severity of gall-stone ileus is due partly to coincident obesity and myocardial disease, but partly also to delay in diagnosis, the obstruction being mistaken at first for a mere repetition of the attacks of cholecystitis from which the patient has in the past been accustomed to recover spontaneously.

PART II

TUMOURS OF THE SMALL INTESTINE

1. AETIOLOGY

196.] Perhaps even less is known of the pathogenesis of tumours of the small intestine than is known of tumour formation in general, and the rarity of *Rarity*

tumours in this situation has never been explained. Cancer of the small intestine is eighty or ninety times absolutely more rare than cancer of the stomach (Rankin and Mayo, 1930), and, square inch for square inch, hundreds of times less common. There is a difference, too, not only in incidence but in age incidence. The mean age incidence of simple tumours of the small intestine is 38, and half of the patients are in their thirties (Rankin and Newell, 1933); the mean age incidence of cancer of the small intestine is 47·5—10 years higher than that of the simple tumours, and 10 years lower than the mean age incidence of cancer of the colon. In Rankin's Mayo Clinic series males are affected as often as females, but in longer collected series (Cameron, 1938) males have suffered more frequently.

Age incidence

Sex incidence

Malignant change in simple tumours

Carcinoma of the small intestine appears sometimes to develop in a pre-existent simple tumour or pancreatic rest. Simpson-Smith (1938) found a relatively high incidence of sarcoma of the intestine in young infants.

2. CLASSIFICATION

Tumours of the small intestine, as of other organs, are classified primarily as benign or malignant.

(1) Benign tumours

A large and varied list could be submitted of benign tumours which, like the cystadenoma and the osteochondroma, have occurred once or twice in the small intestine, and all varieties are relatively rare. Among the less rare are the adenoma, the leiomyoma, the fibroma, the neurofibroma, the lipoma, the haemangioma and the solitary lymphoma. Mucosal rests of heterotopic pancreatic tissue, though not neoplastic, sometimes project into the lumen sufficiently to behave as benign tumours. Carcinoid tumours occasionally have malignant characters, more often in the small intestine than in the appendix, but the majority behave in a benign way.

Mucosal rests

(2) Malignant tumours

Of malignant tumours, both sarcoma and carcinoma may occur in the small intestine. In the sarcoma group, leiomyosarcomatous and undifferentiated forms occur, and conveniently included in this category for surgical purposes are certain "lymphoid tumours" (Warren and Lulenski, 1942), diseases of the local intestinal reticulo-endothelial tissue—lymphosarcoma, reticulum-cell sarcoma and lymphadenoma Two forms of carcinoma occur in the small intestine—the adenocarcinoma and the malignant carcinoids.

Sarcoma

3. MORBID ANATOMY

The *adenoma*, least rare of the simple tumours, is single four times more often than multiple, pedunculated ten times more often than sessile; it forms a smooth projection into the lumen and is said to be liable to malignant change. The *leiomyoma* is always single, taking the form of a smooth and usually sessile projection from the mucous or serous surface; it may undergo a sarcomatous change (Golden and Stout, 1941). The *fibroma* may originate in any layer as a hard, white, whorled tumour; always single, it usually

includes areas of hyaline, myxomatous or calcareous degeneration. The *neurofibroma* presents similar characters to the naked eye (Fig. 44). The *lipoma* occurs as a soft yellowish tumour, intramural, submucous or subserous, and the *haemangioma*, occurring also in any layer, does not differ from haemangiomas in other sites; occasionally a diffuse haemangiomatosis of the small intestine, or even of the whole gastro-intestinal tract has been described, and the multiple hereditary telangiectases of Osler may occur with the same tendency to haemorrhage in the intestine as in such situations as skin and nasal mucosa. The solitary *lymphoma* gives a white, encapsulated, submucous tumour. *Accessory pancreatic tissue* may occur in the small intestine as a pale mucosal plaque slightly elevated and a centimetre or so in diameter, closely resembling a simple tumour in its appearance and its effects. It is said to be the seat sometimes of malignant change.

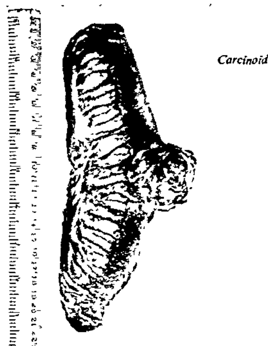


FIG 44.—Subserous tumour of small intestine, nature not obvious from inspection. Proved histologically to be *neurofibroma*.

Carcinoid

The *carcinoid* tumour of the small intestine, like that of the appendix, is usually small, circumscribed, globose or ovoid, lemon-yellow in colour and situated in or apparently just under the mucosa (*see* Appendix—Tumours of). It resembles the carcinoid too in its histological appearance, though it is more cellular and contains fewer nerve fibres (Porter and Whelan, 1939); it exhibits the same affinity for silver salts and has been similarly entitled "chromaffinoma" or "argentaffinoma". Its origin is equally debated. Its commonest site in the small intestine is the ileum, but the tumour has occurred also in the jejunum and even in a Meckel's diverticulum. Most carcinoids are symptomless and discovered only after death. Those carcinoids which have produced symptoms have occurred at a later age than carcinoid of the appendix (Humphreys, 1934). They are multiple in perhaps as many as 50 per cent of cases (Dockerty and Ashburn, 1943). Of those patients who undergo operation, from 20 to 40 per cent have glandular metastases, whereas less than 5 per cent of appendix carcinoids metastasize. There is some reason to regard the carcinoid of the small intestine as a low-grade carcinoma rather than as a benign tumour.

Sarcoma

The *sarcomas* in general, primitive round-cell, spindle-cell, or leiomyosarcoma, tend to form bulky tumours (Simpson-Smith, 1938), though they do sometimes occur in annular form The lymphoid and reticular sarcomas usually convert the bowel into a hard, rigid, thick-walled tube. Any of these forms may project from the serous or mucous surface, or infiltrate the muscle layers (subserous, submucous or intramural forms).

Carcinoma

Adenocarcinoma may arise as a primarily malignant tumour, or by malignant degeneration in an adenoma or a pancreatic rest. It seems commoner in the ileum than in the jejunum (Fraser, 1945), though in the Mayo Clinic series there were 31 jejunal to 21 ileal cases (Mayo, 1940). It gives rise usually to an annular stricture, less commonly to a fungoid growth. Metastases are often general when the primary growth is still small.

4. CLINICAL PICTURE

Obstructive signs late

Since the contents of the small intestine are fluid, obstructive signs are late, and seldom due to simple mechanical occlusion by tumour.

Benign tumours

Haemorrhage

Intussusception

Simple tumours may, rarely, ulcerate and give rise to painless melaena, and the angioma and the leiomyoma too, have on occasion led to rapid unconsciousness and even death from bleeding (Cave, 1932); but benign tumours usually pass unnoticed unless they lead to intussusception or, rarely, and in pedunculated tumours, to volvulus. Most intussusceptions in adults, and many enteric intussusceptions in childhood, are due to benign tumours. A bowel loop bearing a pedunculated subserous tumour may, less commonly, undergo volvulus, and occasionally a tumour may occlude by kinking or by the formation of adhesions (Douglas, 1922). The sarcomas, if the primary growth gives rise to symptoms, have effects similar to those of benign tumours, but usually distant metastases are present before the primary disease is detected.

Sarcoma

Carcinoma

The carcinomas and the lymphoid tumours (Cameron, 1938), although they may ulcerate and give rise to haemorrhage, or fungate as palpable tumours, usually encircle the bowel to form annular strictures, but here, as in sarcoma, metastases are sometimes general whereas the primary tumour is still small and symptomless.

Perforation rare

Perforation of a tumour of the small intestine occurs very rarely (Lewis, 1939).

Radiological investigation

Contra-indications

It is seldom possible, even by an elaborate technique, to demonstrate tumours of the small intestine radiologically. Indeed, it is doubtful if a standard opaque meal should be given to patients suffering from a developing obstruction lest occlusion be made complete by the barium. The tumour is in any case seldom outlined, though Lingley (1936) was able to demonstrate the cavity of a malignant enteric ulcer filled by a residue of barium after the remainder of the bowel had emptied.

Screening for an hour

Several special radiological methods have been designed for the detection of tumours of the small intestine. Akerlund (1932) advised screening for an hour after a meal of barium in aqueous suspension, and illustrated 4 cases in which the diagnosis was undoubtedly made by the observation of obliteration of rugae, stiffness of a coil, tenderness, or a filling defect. Mayo and Nettrour (1936) made a correct diagnosis in 10 of 31 patients studied by this method, and Mitchell-Heggs (1937) has also used it with success.

Injection along Miller-Abbott tube

Simpson-Smith (1938) advised that when obstruction is present, but neither complete nor acute, a Miller-Abbott tube be passed till it meets the obstruction; barium injected along the tube outlines the tumour. Frank, Miller and Bell (1942) record the advantage of Simpson-Smith's method.

5. TREATMENT

A benign tumour detected at operation and complicated neither by intussusception nor by acute obstruction may be resected with that portion of the bowel wall to which it is attached, the resultant defect being closed transversely: if the attachment is broad a whole segment may have to be resected, and continuity re-established by anastomosis. If a simple tumour is found at the apex of an intussusception, it may be similarly dealt with, though in this case a considerable length of bowel may require resection with the tumour if intussusception has threatened its viability. It is sometimes difficult to distinguish by palpation a simple tumour at the apex of an intussusception from the oedematous apex of what has been a primary intussusception. If there is doubt, the bowel may be opened and the apex directly inspected. A simple tumour which has caused mechanical obstruction may be resected, or, if the patient's general condition is serious, a lateral anastomosis may be performed immediately, and resection postponed to a later date (Mitchell-Heggs, 1939). Jejunostomy should be avoided; its mortality, admittedly at a time before the importance of replacement of lost fluid was understood, was 75 per cent (d'Allaines, 1929).

Benign tumours

The treatment of malignant tumours of the small intestine is wide resection of the tumour-bearing segment.

6. PROGNOSIS

The prognosis of benign tumour treated by excision is favourable Although 20–40 per cent of patients with carcinoids have metastases already at the time of operation and do not survive for long, a very large majority of those who have a single primary carcinoid removed enjoy a complete recovery. The prognosis in carcinoma of the small intestine is poor—the fatality rate after operative removal is 30 per cent, and only 10 per cent survive for 5 years (Cameron, 1938). Lymphosarcoma seems to follow a less rapidly fatal course in the small intestine than elsewhere: in Cope and Grant's (1942) case, the patient had suffered from abdominal symptoms for 6 years before operation; of 85 cases of intestinal lymphosarcoma collected by Ullman and Abeshouse (1932) 55 patients survived for 2 years, and 8 for at least 7 years, and Cameron (1938) records survival for 8, 13 and 20 years after removal of lymphosarcomas of the bowel.

Benign tumours

Carcinoids

Carcinoma

Lymphosarcoma

PART III
TUBERCULOSIS OF THE INTESTINE

1. VARIETIES

Ulcerative and hyperplastic forms

197.] Tuberculous disease occurs in the intestine in two distinct forms—the ulcerative and the hyperplastic. The former is usually a secondary form of tuberculosis in a sanatorium patient whose resistance is low in relation to the virulence of the infection, and who suffers primarily from an active and open pulmonary lesion; the hyperplastic form appears to be due to a relatively attenuated and often apparently primary infection of a patient whose resistance is high.

2. BACTERIOLOGY AND ROUTE OF INFECTION

Human and bovine infection

The ulcerative form of the disease is due usually to infection by the human bacillus, though the bovine form may occur in the ulcerative tuberculous enteritis of young children. In hyperplastic intestinal tuberculosis the bovine organism is usually blamed, though the disease has occurred in Japanese districts in which cow's milk is unknown.

Ingestion

In both forms the infection enters the intestinal wall from the lumen. In the ulcerative form the bacilli are swallowed in infected sputum, in the hyperplastic form they are ingested with milk or other food. In both cases the organisms are said to enter the bowel wall by way of a solitary lymph follicle or Peyer's patch, usually of the lower ileum. It should be added, however, that intestinal tuberculosis has been reproduced experimentally in susceptible animals not only by feeding with virulent bacilli but also by intravenous inoculation.

Portal of entry

3. MORBID ANATOMY

(1) Ulcerative form

[illegible] and [illegible]

covered by weak granulations and [illegible] and sometimes caseated. Perforation seldom occurs, but the ulcers may heal with stricture formation, to produce intestinal obstruction later.

(2) Hyperplastic form

The hyperplastic variety may [illegible] colon or, rarely, a still lower segment of [illegible] stics are those not so much of inflammation as of repair. The [illegible] opic

tubercles, first in submucosa and later in subserosa, are strangled in a mass of fibro-fatty tissue which converts the bowel into a thick-walled, rigid tube; it is this violent local healing reaction, rather than the serious nature of the infection, which produces the pathological effects of the disease. If the disease affects the ileo-caecal region, as it usually does, the appendix may be buried in the reparative mass. A few tubercles may be visible upon the serous aspect, and the omentum may be wrapped round and adherent to the diseased bowel, but there is seldom any caseation, cold abscess or sinus formation. The mucosa is thrown into valvular or polypoid folds, oedematous or rigidly fibrous, and there may be some superficial ulceration, not necessarily tuberculous in appearance. The ileo-caecal valve, if the disease is located near it, is usually shrunken, folded and distorted. The lumen is narrowed by the fibrous constriction of the wall of the bowel and by the heaping together of mucosal folds; sometimes occlusion is rendered complete by the impaction of a fruit stone or other foreign body or of a faecolith, at the site of narrowing. The affected segment of the colon is shortened; a tuberculous ascending colon, for example, may by its contraction draw up the caecum so that the ileum rises to enter the caecum at an angle of 180 degrees. The regional lymph glands may be enlarged, oedematous, sprinkled with tubercles, frankly caseous or calcified, and sometimes the adjacent peritoneum of mesentery or parietes is congested or studded with tubercles. It is very rare for hyperplastic tuberculosis to affect multiple levels of intestine, but sometimes one or more simple tuberculous strictures, apparently the result of antecedent ulceration, may be found at a higher level in the small intestine.

Submucosa and subserosa

Serosa and mucosa

Ileo-caecal valve

Lumen

Lymph glands

Coincident tuberculous strictures

Histologically there is a diffuse fibrous reaction, with few giant cells, little caseation and rarely tubercles of typical pattern. A confident histological diagnosis is more readily based upon examination of a lymph gland than upon study of the affected bowel wall. It is difficult to demonstrate acid-fast bacilli in sections of the fibro-fatty mass, and guinea-pig inoculation may be negative. Hypertrophic tuberculosis bears a close similarity to the chronic form of regional ileitis (Crohn's disease) and the two conditions are not infrequently confused. Hypertrophic tuberculosis has become extremely uncommon since Crohn's description of regional ileitis. A diagnosis of hypertrophic tuberculosis is justified only if guinea-pig inoculation is positive, or if acid-fast bacilli are detected in sections, or if typical tubercles are observed by the pathologist in the intestinal lesion or in one of the related lymph glands.

4. CLINICAL PICTURE

(1) Ulcerative form

The ulcerative variety gives rise to the signs of enteritis in sanatorium patients, and a few of these ulcers, healing with stricture, occasion acute obstruction later.

(2) Hyperplastic form

The hyperplastic form, which is commonest between 20 and 40 years of age, and affects females more often than males, gives rise in the first place to the symptoms of increasing chronic intestinal obstruction—vague dyspepsia, a feeling of distension most noticeable after a meal and attacks of abdominal colic. These may be accompanied by a dull pain in the right iliac fossa.

Age and sex

Chronic intestinal obstruction

1. VARIETIES

Ulcerative and hyperplastic forms

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Portal of entry

3. MORBID ANATOMY

(1) Ulcerative form

The ulcerative variety takes the form of multiple ileal ulcers, oval in shape and circumferentially placed. The edges are grey and undermined, the base is covered by weak granulations and the regional lymph glands are enlarged and sometimes caseated. Perforation seldom occurs, but the ulcers may heal with stricture formation, to produce intestinal obstruction later.

(2) Hyperplastic form

The hyperplastic variety may affect ileum, caecum or ascending colon or, rarely, a still lower segment of large bowel; its pathological characteristics are those not so much of inflammation as of repair. The initial microscopic

tubercles, first in submucosa and later in subserosa, are strangled in a mass of fibro-fatty tissue which converts the bowel into a thick-walled, rigid tube; it is this violent local healing reaction, rather than the serious nature of the infection, which produces the pathological effects of the disease. If the disease affects the ileo-caecal region, as it usually does, the appendix may be buried in the reparative mass. A few tubercles may be visible upon the serous aspect, and the omentum may be wrapped round and adherent to the diseased bowel, but there is seldom any caseation, cold abscess or sinus formation. The mucosa is thrown into valvular or polypoid folds, oedematous or rigidly fibrous, and there may be some superficial ulceration, not necessarily tuberculous in appearance. The ileo-caecal valve, if the disease is located near it, is usually shrunken, folded and distorted. The lumen is narrowed by the fibrous constriction of the wall of the bowel and by the heaping together of mucosal folds; sometimes occlusion is rendered complete by the impaction of a fruit stone or other foreign body or of a faecolith, at the site of narrowing. The affected segment of the colon is shortened; a tuberculous ascending colon, for example, may by its contraction draw up the caecum so that the ileum rises to enter the caecum at an angle of 180 degrees. The regional lymph glands may be enlarged, oedematous, sprinkled with tubercles, frankly caseous or calcified, and sometimes the adjacent peritoneum of mesentery or parietes is congested or studded with tubercles It is very rare for hyperplastic tuberculosis to affect multiple levels of intestine, but sometimes one or more simple tuberculous strictures, apparently the result of antecedent ulceration, may be found at a higher level in the small intestine.

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Age and sex

Chronic intestinal obstruction

Ulceration is exceptional, and diarrhoea and melaena are therefore not common clinical features. Constipation increases until finally a complete obstruction, with vomiting and progressive central abdominal distension, may gradually or suddenly supervene. The general effects of tuberculosis—loss of weight, occasional pyrexia, and sudomotor activity—are not usually prominent.

Acute intestinal obstruction

Clinical examination

On examination, distension and ladder pattern are seen only in the later stages, but right iliac fossa tenderness can usually be elicited and a hard mass can be felt in the ileo-caecal region. Tubercle bacilli cannot be obtained from stool or sputum, and other foci of tuberculosis, in lungs or cervical glands, cannot usually be demonstrated.

Skiagram

Opaque meal or enema

Screening

A straight skiagram of the abdomen may show gas-distended coils of small intestine, with or without multiple fluid levels; the shadows of calcified mesenteric glands are not necessarily significant. Barium meal or enema demonstrates a long regular filling defect, narrowing the lumen to a fine cord which cannot readily be emptied, under the screen, by the pressure of the examining hand. The ascending colon is short and the caecum is seen to be at a high level, the terminal ileum rising vertically to enter it. An opaque enema is preferable to an opaque meal if obstructive signs are present—barium taken by mouth may precipitate an acute obstruction.

5. DIAGNOSIS

The diagnosis of the ulcerative form of tuberculosis of the intestine offers no difficulty—the occurrence of gastro-enteritis in a patient with demonstrable pulmonary tuberculosis can hardly be due to anything else.

The diagnosis in hyperplastic tuberculosis is seldom certain before operation, but a chronic low ileal obstruction in a young adult who presents a firm swelling in the right iliac fossa, and the radiological signs described in the previous paragraph, may be suspected of being due to hypertrophic ileo-caecal tuberculosis.

6. DIFFERENTIAL DIAGNOSIS

Cancer of caecum

Cancer of the caecum or ascending colon may give the same symptoms and signs as hyperplastic tuberculosis, and the age periods of incidence of the two conditions overlap, but in the opaque enema cancer gives a localized and irregular filling defect and usually the ileum is demonstrably unaffected; occult blood is found in the stool more often in cancer than in hypertrophic tuberculosis.

Actinomycosis

Actinomycosis presents nowadays usually as a sinus persisting after appendicectomy—sinus is unusual in hypertrophic tuberculosis. Ileo-caecal actinomycosis, in the form of abdominal swelling without sinus formation, is difficult to distinguish from hypertrophic tuberculosis unless it is accompanied by disease in liver or lung, or unless it has invaded the psoas to cause fixed flexion of the hip.

Regional ileitis

Regional ileitis in its chronic obstructive form cannot be distinguished from hypertrophic tuberculosis before operation; the shadows of tuberculous glands in the straight skiagram give weight to a clinical diagnosis of tuberculosis. At operation it is almost as difficult to distinguish the conditions as

it is before operation, unless tubercles are visible upon the surface of the bowel, the mesentery or the regional lymph glands. The specimen removed by resection may still satisfy either diagnosis equally well, and the histological appearances may not be conclusive, unless definite tubercles and giant-cell systems are observed in bowel lesion or gland.

When the patient presents with acute symptoms of vomiting, distension and a palpable tumour in the right iliac fossa, the resemblance to acute appendicitis with abscess may be close. There is, however, no leucocytosis in hyperplastic tuberculosis, however acute the symptoms; when a palpable abscess has supervened in acute appendicitis the white-cell count is always high.

Appendicitis

When hyperplastic tuberculosis affects the pelvic colon it may simulate either cancer or peridiverticulitis, but the opaque enema serves usually to distinguish these; sigmoidoscopy, by proving the type of ulceration present, is particularly valuable in excluding or establishing a diagnosis of cancer. Actinomycosis and Crohn's disease may occur also at this site.

Tuberculosis of pelvic colon

7. PROGNOSIS

(1) Ulcerative form

The prognosis in ulcerative tuberculosis of the intestine is bad. It is frequently a terminal phenomenon in the sanatorium patient. Its progress, however, often reflects the activity of the pulmonary lesion—if the lung recovers, the intestinal ulcers heal.

(2) Hyperplastic form

The prognosis in the hyperplastic form of the disease is good, provided surgical treatment is adequate. The patient in this disease has a high degree of resistance, other active foci are absent, and life and health are threatened chiefly by the mechanical effects of repair.

8. TREATMENT

(1) Ulcerative form

The treatment of tuberculous ulceration in its acute stage does not concern the surgeon. If the ulcers heal with stenosis, however, the patient may come to the surgeon later for relief of acute or chronic intestinal obstruction. The occlusion should be relieved by lateral anastomosis, but it should be remembered at operation that the strictures are often multiple and more than one anastomosis may be required.

Lateral anastomosis

(2) Hyperplastic form

The treatment of the hyperplastic form of tuberculosis is operative. In most circumstances, resection of the hyperplastic segment, by right hemicolectomy for example in the ileo-caecal form of the disease, should be performed. A short-circuit exclusion alone, while sometimes followed by permanent relief of symptoms, and even on occasion by progressive reduction in size of a palpable tumour, may not completely alleviate symptoms, and leaves two blind loops, proximal and distal respectively to the occluding disease, between the disease and the anastomosis.

Right hemicolectomy

If obstruction is not complete, the hemicolectomy may be performed conveniently and safely in one stage. If operation is performed in the presence of

obstruction, the obstruction should be relieved by lateral anastomosis—ileo-transverse colostomy in the case of ileo-caecal disease—and resection postponed until the patient has recovered from the effects of the obstruction, or even until it is clear that pain has not been relieved, that swelling is increasing in extent or, by radiological or clinical examination, that the proximal or distal loop is dilating between disease and anastomosis.

Search for concomitant strictures

When operation is performed, a search should be made for concomitant tuberculous strictures higher in the small intestine; a stricture affecting a segment of bowel adjacent to the hyperplastic disease may be included in the resection of the main disease; more distant strictures may require one or more entero-anastomoses for the obstruction which they occasion or threaten.

PART IV

REGIONAL ILEITIS (CROHN'S DISEASE; NON-SPECIFIC GRANULOMA)

1. DEFINITION

198.] Regional ileitis is a non-specific granuloma of the small intestine; a similar inflammatory state may occur in other hollow viscera.

2. NOMENCLATURE

Regional ileitis

The term "regional ileitis" has been chosen to head this account since it is probably the title most commonly used; the title is not a happy one, however, for it is not certain that the disease is inflammatory, other organs besides the ileum may suffer, and other conditions, more specifically inflammatory, may affect the ileum in a regional way. "Crohn's disease" is convenient in avoiding any aetiological connotation and, though unwelcome to those who dislike

Crohn's disease

eponymous nomenclature, is not applied without some justice. Crohn did not first describe the condition, but he clearly classified its symptoms and its stages and only since his description has the condition been widely recognized as a clinical entity. Koch (1903) describing the condition in the colon, styled it "inflammatory fibrous colonic tumour", whereas Wilensky and Moschowitz (1927) applied the title "non-specific granuloma"; this last seems logically the least objectionable name for it, and will be used throughout this article.

Non-specific granuloma

3. HISTORY

Abercrombie (1828) described a condition which seems to have been non-specific granuloma. Virchow (1853) was familiar with the lesion, ascribing it to chronic peritonitis, and Koch (1903) reported its occurrence in the colon. Robson (1908) clearly distinguished between cancer and inflammatory tumours of gullet, stomach, small intestine, colon and rectum, but probably included in the last situations cases of diverticulitis of the colon and lymphogranulomatous stricture of the rectum. To Wilensky and Moschowitz (1927) must be given credit for the first clear description of the condition and its separation from other inflammatory lesions. Crohn, Ginzburg and Oppenheimer (1932) distinguished the various pathological types and related them to the clinical manifestations of the disease.

4. MORBID ANATOMY

Sites

Non-specific granuloma is commonest in the ileum but has been described in the colon, jejunum, rectum, duodenum, oesophagus and stomach in order of diminishing frequency. Two or more areas may be simultaneously affected, usually ileum and colon (Colp, 1934); Dalziel (1913) recorded a case which seems to have been non-specific granuloma and which involved the whole gastro-intestinal tract.

Crohn's classification

Four phases of the disease were distinguished by Crohn, Ginzburg and Oppenheimer (1932), and Crohn's classification will be closely adhered to here, though the relation of each phase to the others is not yet clearly understood; one or more stages may seem to be lacking in individual cases, sometimes they develop in an atypical order and sometimes they occur together in varying combinations.

(1) Acute ulcerative phase

The first or acute ulcerative phase resembles any acute non-specific ulcerative enteritis—Harris, Bell and Brunn (1933) described a small oval ulcer on the mesenteric border of the ileum as the primary lesion, and Henke and Lubarsch (1938) regard such an ulcer as a portal of entry for infection, though this is still conjectural and the temporal priority of an ulcer is not yet substantiated; in hyperplastic tuberculosis, the pathology of which is similar, frank ulceration of the mucosa, when it is present, is not necessarily tuberculous—it may be secondary to fibrous hyperplasia in the submucosa.

(2) Acute phlegmonous phase

The second form or phase is acute phlegmonous enteritis; this seems sometimes to precede ulceration or to develop and continue without ulceration.

The serous surface of the affected segment is bright red in colour, with congested vessels, and sometimes a covering of fibrinous exudate in sheets or shreds, or of adherent omentum. The whole bowel wall is thick with an oedema which affects chiefly the submucosa and subserosa. The mucosa, which may or may not be ulcerated, is thrown into oedematous folds. Perforation, followed by localized abscess or fulminating peritonitis, has complicated this stage of the disease. Histologically (Schepers, 1945) the thick bowel wall shows an oedema and lymphangiectasis of submucosa and subserosa, which is soon infiltrated by lymphocytes; these in turn are replaced by proliferating histiocytes. The argentophil fibrillary processes of these histiocytes form a reticulum in which fibroblasts build the fibrous tissue of the succeeding fibrous stage.

Histology

(3) Chronic hyperplastic phase

In the fibrous or hyperplastic phase of regional enteritis the pathological appearances are difficult to distinguish from those of hyperplastic ileo-caecal tuberculosis. The affected segment is thickened by submucous and subserous deposits of fibro-fatty tissue, which may contain sparse giant cells of foreign-body type. The lumen is narrowed by thickening of the bowel wall, and the mucosa is thrown into folds and sometimes ulcerated.

(4) Chronic phase of abscesses and fistulae

The untreated disease may or may not proceed to the final chronic phase of abscess and fistula formation. External faecal fistulae may occur, or internal fistulae may be established with the bladder or with other portions of the gastro-intestinal tract. The fistulae are preceded by suppuration round a granulomatous bowel loop, in the phlegmonous or in the hyperplastic phase, which has developed adhesions with the parietal peritoneum. Most external fistulae arise as sinuses persisting after appendicectomy (Wilensky, 1939) or after anastomosis or incomplete removal of affected bowel (Clark and Dixon, 1939). A few external and most internal fistulae arise spontaneously.

Enlarged regional lymph glands

Whatever the phase of the disease in the intestine, the regional lymph glands are usually enlarged, being the seat of an acute inflammatory reaction in the early stages of the disease and a reticulum-cell hyperplasia in the later stages. The mesentery and parietal peritoneum may be thickened and congested, and there may be a small amount of free fluid in the peritoneal cavity.

5. AETIOLOGY AND PATHOGENESIS

Claim to rank as distinct entity

Non-specific granuloma has a high though not fully proven claim to rank as a distinct pathological entity, and it is convenient to regard it as a single and distinct disease until its cause or causes are known. The pathological appearances of the individual phases are very constant, although their relation to each other is variable, and the serial passage of phase into phase has not always been demonstrable. The close resemblance of each phase to some other recognized and independent disease long delayed the recognition of non-specific granuloma as a distinct pathological process in its own right, and, conversely, has perhaps led to recent inclusion of cases of these other diseases in the category of non-specific granuloma. The acute ulcerative

Resemblance to other diseases

phase, for example, does not differ from other forms of ulcerative enteritis, and cannot be recognized as non-specific granuloma unless it proceeds to or is associated with one of the other characteristic phases. The acute phlegmonous phase is the most characteristic of the four, but in its pathology it is identical with the acute phlegmonous enteritis which may follow impaction of a sharp foreign body in the lumen of the intestine; it also closely resembles the acute phlegmonous gastritis of Varandeus and Cruveilhier, a streptococcal intramural infection of the stomach from a local portal of entry through broken mucosa in gastric disease, or by the blood stream in puerperal and other fevers: Ralphs (1938) regards the disease as a non-specific intramural inflammation occurring in response to any infection which penetrates the bowel wall. The close similarity between the chronic hyperplastic phase and hyperplastic ileo-caecal tuberculosis has already been noted—indeed Taylor (1945) recorded a case in which the bowel lesion presented the appearance of a regional ileitis and could not be proved to be tuberculous, though the enlarged regional lymph glands were typically tuberculous and furnished acid-fast bacilli. The fourth phase of abscesses and fistulae resembles actinomycosis, but the enlargement of the regional lymph glands is sufficient in itself to absolve actinomycosis from all blame. This phase also bears a pathological resemblance to lymphogranuloma inguinale as it occurs in the rectum, but in patients suffering from non-specific granuloma the Frei test is negative (Koster, Kasman and Sheinfeld, 1936).

Inflammatory process

The cause of non-specific granuloma is not known. All its phases suggest an inflammatory process, and its onset may be preceded by upper respiratory tract infection, but cultures of bowel wall, mesentery and regional lymph glands have seldom yielded any growth (Mock, 1931; Pumphrey, 1938), though in one of Mailer's cases a *Streptococcus viridans* was grown from the blood (Mailer, 1938). The specific organism found by Brust and Bargen (1934) is regarded by most bacteriologists as a secondary invader, as is the streptococcus isolated by Jackman (1934). Felsen (1936) obtained, in some cases, a positive agglutination reaction to organisms of the dysenteric group, but no other worker has shown that the incidence of positive dysenteric agglutination is higher in granuloma patients than in unaffected subjects of the same geographical district. The close resemblance of the chronic phase to hyperplastic ileo-caecal tuberculosis led Dalziel (1913) and, later, W. J. Mayo (quoted by Clark and Dixon, 1939) to suggest that some variant of the tubercle bacillus may be responsible—for example, that which causes Johne's disease of cattle (M'Fadyean, 1907). Since no bacterium can reasonably be blamed, it is suggested that a virus may be responsible for so apparently inflammatory a disease (Koster, Kasman and Sheinfeld, 1936); this theory is supported by the close similarity of the histological features to those of lymphogranuloma inguinale (granulomatous stricture of the rectum) which is a virus infection, and by the significant frequency with which non-specific granuloma is preceded by upper respiratory tract infection. Wilensky (1939) has related non-specific granuloma to the non-specific mesenteric lymphadenitis of children, and ascribes both to virus infection. The lymph-gland enlargement is similar in the two conditions (Lick, 1938), and in mesenteric lymphadenitis a red flush and oedema are sometimes observed in the adjacent intestine. "The lymph-gland enlargement of granuloma *is* a non-specific lymphadenitis, and

Bacteriology

Relation to tuberculosis

Virus

Relation to mesenteric adenitis

the inflammation sometimes observed in the bowel in a case of lymphadenitis *is* a regional ileitis." (Aird, 1945.)

Not all observers agree that non-specific granuloma is an infection of the bowel. Upham (1938), stressing the lymphangiectasis and oedema of early cases, suggests that infection, entering the ileal lymphatics at the ileo-caecal angle, may lead to lymphadenitis and lymphatic obstruction, and that this in turn may lead to oedema of the bowel and formation of the granuloma. Reichert and Mathes (1936) blame a combination of infection and lymphatic obstruction, and have produced enteric lymphoedema in animals by the injection of sclerosing solutions into the mesenteric lymphatic channels. Taylor's case of non-specific granuloma associated with definitely tuberculous mesenteric adenitis supports this contention (Taylor, 1945). Other authors have suggested an allergic vasodilatation of the intestinal arteries similar to that which occurs in Henoch's colonic purpura, and the vivid red of the "phlegmonous stage" lends colour to this view. Hadfield and Garrod (1942) regard non-specific granuloma as a local intestinal reticulosis, and adduce in evidence the lymphocytosis and reticulum-cell hyperplasia which precedes fibroblastic activity.

Lymph stasis

Allergy

Reticulosis

The condition is, however, so inflammatory in appearance at operation that it is difficult for the surgeon not to believe that it is an inflammation until some other cause is proved; since bacteria cannot usually be isolated, the infecting agent may be presumed to be a virus.

6. CLINICAL PICTURE

Sex and age incidence

The sexes are equally affected. The disease is commonest in early adult life, but Ebrill (1945) has recorded it in a child of 9, and Pratt and Simpson (1942) in a child of 7. Six of Koster's 67 patients were less than 10 years of age, and 5 were over 70 years.

(1) Acute ulcerative phase

The acute ulcerative phase cannot be distinguished from other forms of gastro-enteritis unless a later phase coexists.

(2) Acute phlegmonous phase

In the acute phlegmonous phase general abdominal colic and sometimes vomiting and pain in the right iliac fossa are present. At this stage the patient is usually constipated but there may be a history of previous diarrhoea with melaena There has sometimes been a recent upper respiratory tract infection. There is tenderness, but not extreme tenderness, and often a swelling vaguely palpable in the right lower quadrant, and if the swelling is present it can sometimes be moved by the palpating hand. There is little rigidity and it is often appreciated that the swelling is less hard than is an appendix abscess There is little or no fever or leucocytosis in most cases.

(3) Chronic hyperplastic phase

The chronic hyperplastic phase of the disease manifests the symptoms and signs of progressive intestinal obstruction seldom proceeding to complete occlusion. A history of previous diarrhoea, of melaena or of an acute attack of abdominal colic with pain in the right iliac fossa can frequently be elicited.

History

A firm mass is freely palpable too in the right lower quadrant, with relatively little muscular rigidity over it, and such a mass lying in the pouch of Douglas may be felt from the rectum. The swelling may still be mobile. X-ray examination is helpful in the detection of this stage of the disease. The barium meal shows loss of segmentation and peristalsis in the lower ileum, and a lumen of uniform diameter persists in spite of manual manipulation. In later stages barium in the lower ileum tapers to a fine cord before entering the caecum—this is the "string sign" of Kantor (1934). Weber (1938) suggests that a barium enema should precede the opaque meal; an ileal lesion can often be outlined if the coils of small intestine are manipulated upwards out of the pelvis.

X-ray examination

(4) Chronic phase of abscesses and fistulae

In the phase of chronic abscesses and fistulae a history may be obtained of the symptoms of preceding phases; the tumour mass is usually clearly palpable, and the radiographic signs described in the previous paragraph can usually be observed. Internal communications are sometimes outlined by barium meal, even if such symptoms as true faecal vomiting or diarrhoea with the passage of undigested food do not suggest an entero-gastric fistula, or cystitis or pneumaturia an entero-vesical communication. External fistulae may be outlined by fistulography: some abscesses extend very widely, tracking upwards to reach the subdiaphragmatic region, laterally to open in the loin, or even downwards through the pelvic floor to reach the ischio-rectal fossa as one form of pelvi-rectal fistula.

Fistulography

7. DIFFERENTIAL DIAGNOSIS

(1) Acute ulcerative phase

The initial acute ulcerative phase can be differentiated from other varieties of gastro-enteritis only if other phases coexist. The acute phlegmonous stage is usually confused with acute appendicitis, but should be detected clinically if there has been diarrhoea and melaena and if a swelling of softer consistence than that of an appendix mass is palpable.

(2) Chronic proliferative phase

The chronic proliferative phase is to be radiologically differentiated from chronic hyperplastic ileo-caecal tuberculosis by the predominant involvement of ileum which is usual in non-specific granuloma, and by the normal girth, outline and length of the right colon, which is predominantly affected by ileo-caecal tuberculosis of the hyperplastic type. Cancer of the caecum presents on palpation a harder and more irregular tumour than either hyperplastic tuberculosis or non-specific granuloma, and in the skiagram a more localized and irregular filling defect than either of these diseases: it affects the caecum or colon and leaves the ileum unaffected.

Differential diagnosis

(3) Chronic phase with abscesses and fistulae

The chronic granuloma with abscess and fistula formation may be confused with any form of chronic intra-abdominal suppuration, and an abscess may attract attention by pointing in abdominal, lumbar, subphrenic, pelvic or even ischio-rectal regions. A granulomatous fistula occurring as a persistent sinus after appendicectomy must be distinguished from actinomycosis and

the inflammation sometimes observed in the bowel in a case of lymphadenitis *is* a regional ileitis." (Aird, 1945.)

Not all observers agree that non-specific granuloma is an infection of the bowel. Upham (1938), stressing the lymphangiectasis and oedema of early cases, suggests that infection, entering the ileal lymphatics at the ileo-caecal angle, may lead to lymphadenitis and lymphatic obstruction, and that this in turn may lead to oedema of the bowel and formation of the granuloma. Reichert and Mathes (1936) blame a combination of infection and lymphatic obstruction, and have produced enteric lymphoedema in animals by the injection of sclerosing solutions into the mesenteric lymphatic channels. Taylor's case of non-specific granuloma associated with definitely tuberculous mesenteric adenitis supports this contention (Taylor, 1945). Other authors have suggested an allergic vasodilatation of the intestinal arteries similar to that which occurs in Henoch's colonic purpura, and the vivid red of the "phlegmonous stage" lends colour to this view. Hadfield and Garrod (1942) regard non-specific granuloma as a local intestinal reticulosis, and adduce in evidence the lymphocytosis and reticulum-cell hyperplasia which precedes fibroblastic activity.

Lymph stasis

Allergy

Reticulosis

The condition is, however, so inflammatory in appearance at operation that it is difficult for the surgeon not to believe that it is an inflammation until some other cause is proved; since bacteria cannot usually be isolated, the infecting agent may be presumed to be a virus.

6. CLINICAL PICTURE

Sex and age incidence

The sexes are equally affected. The disease is commonest in early adult life, but Ebrill (1945) has recorded it in a child of 9, and Pratt and Simpson (1942) in a child of 7. Six of Koster's 67 patients were less than 10 years of age, and 5 were over 70 years.

(1) Acute ulcerative phase

The acute ulcerative phase cannot be distinguished from other forms of gastro-enteritis unless a later phase coexists.

(2) Acute phlegmonous phase

In the acute phlegmonous phase general abdominal colic and sometimes vomiting and pain in the right iliac fossa are present. At this stage the patient is usually constipated but there may be a history of previous diarrhoea with melaena. There has sometimes been a recent upper respiratory tract infection. There is tenderness, but not extreme tenderness, and often a swelling vaguely palpable in the right lower quadrant, and *if the swelling is present it can sometimes be moved by the palpating hand.* There is little rigidity and it is often appreciated that the swelling is less hard than is an appendix abscess. There is little or no fever or leucocytosis in most cases.

(3) Chronic hyperplastic phase

The chronic hyperplastic phase of the disease manifests the symptoms and signs of progressive intestinal obstruction seldom proceeding to complete occlusion. A history of previous diarrhoea, of melaena or of an acute attack of abdominal colic with pain in the right iliac fossa can frequently be elicited.

History

A firm mass is freely palpable too in the right lower quadrant, with relatively little muscular rigidity over it, and such a mass lying in the pouch of Douglas may be felt from the rectum. The swelling may still be mobile. X-ray examination is helpful in the detection of this stage of the disease. The barium meal shows loss of segmentation and peristalsis in the lower ileum, and a lumen of uniform diameter persists in spite of manual manipulation. In later stages barium in the lower ileum tapers to a fine cord before entering the caecum—this is the "string sign" of Kantor (1934). Weber (1938) suggests that a barium enema should precede the opaque meal; an ileal lesion can often be outlined if the coils of small intestine are manipulated upwards out of the pelvis.

X-ray examination

(4) Chronic phase of abscesses and fistulae

In the phase of chronic abscesses and fistulae a history may be obtained of the symptoms of preceding phases; the tumour mass is usually clearly palpable, and the radiographic signs described in the previous paragraph can usually be observed. Internal communications are sometimes outlined by barium meal, even if such symptoms as true faecal vomiting or diarrhoea with the passage of undigested food do not suggest an entero-gastric fistula, or cystitis or pneumaturia an entero-vesical communication. External fistulae may be outlined by fistulography: some abscesses extend very widely, tracking upwards to reach the subdiaphragmatic region, laterally to open in the loin, or even downwards through the pelvic floor to reach the ischio-rectal fossa as one form of pelvi-rectal fistula.

Fistulography

7. DIFFERENTIAL DIAGNOSIS

(1) Acute ulcerative phase

The initial acute ulcerative phase can be differentiated from other varieties of gastro-enteritis only if other phases coexist. The acute phlegmonous stage is usually confused with acute appendicitis, but should be detected clinically if there has been diarrhoea and melaena and if a swelling of softer consistence than that of an appendix mass is palpable.

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Differential diagnosis

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from fistula associated with a residual foreign body—a surgical instrument, a swab or a large appendicular concretion. The diagnosis of actinomycosis is easy if the causative fungus can be grown in anaerobic culture from sulphur granules in the discharge or from the wall of a sinus; if the removed appendix is available its wall and content may be re-examined for mycelium or rods. Fixed flexion of the thigh, if it is present, is a valuable sign of invasion of the psoas by actinomycosis. Positive radiographic evidence of chronic ileitis may be available if the fistulae are due to that disease.

(4) Extra-ileal non-specific granuloma

Granuloma of the jejunum is seldom diagnosed before operation, though that diagnosis should be considered in a patient whose symptoms and signs are those of granuloma, but whose palpable tumour is in an unusual position (Barrington-Ward and Norrish, 1938). The effects of granuloma of the left colon (James, 1938) are difficult to distinguish from those of diverticulitis by other than radiological methods, unless the patient is a young adult, when granuloma is the more likely clinical diagnosis; if diverticula cannot be seen in the barium enema, however, they may safely be excluded. The long smooth stricture of granuloma of the left colon cannot be mistaken for carcinoma. Non-specific granuloma of the rectum can be distinguished from venereal stricture by the Frei test.

Frei test

8. TREATMENT

Exclusion operation

Though the acute phlegmonous form of non-specific granuloma has been known to resolve without active treatment, it is generally agreed that at least an exclusion operation should be performed by lateral anastomosis, to prevent the development of the chronic form of the disease. Exclusion of the affected segment may not completely relieve the symptoms; Brown and Donald (1942) found recurrence of symptoms in 8 cases of 22 treated by anastomosis alone, and Clark and Dixon (1939) found that toxic symptoms sometimes persist if only an anastomosis is done. On the other hand, Garlock and Crohn (1945) report a happier experience of simple anastomosis—no deaths and only 14 per cent of recurrence after short-circuit alone: it is possible, however, that in this series the more severe local lesions were resected—milder cases resolved after anastomosis. Clark and Dixon (1939) regard one-stage resection as the procedure of choice; they had fewer fatalities after this procedure than after the two-stage operation, in which leakage from blind bowel ends was frequent. Garlock and Crohn (1945) also found one-stage resection more satisfactory than two-stage; in the former, 16 per cent died and in 16 per cent there was recurrence, while after the two-stage procedure 12 per cent died and recurrence followed more than one-third of the operations. These last authors admit, however, that the more severe lesions were likely to be treated by operation in stages.

It is a safe rule to resect in one stage if the patient is not dangerously ill and the bowel not obstructed.

The last phase of the disease, with abscess and fistula formation, is best treated by a complete exclusion of the disease and fistulous segment, and by adequate drainage of abscesses. The fistulous segment may be removed at a second operation if discharge continues.

PART V

CONGENITAL ATRESIA OF THE SMALL INTESTINE

1. DEFINITION

199.] Congenital atresia is an interruption, present at birth, in the continuity of the intestinal lumen.

2. AETIOLOGY

Foetal mechanism unexplained

The foetal mechanism responsible has not been explained. Evidence of a genetic influence is lacking. Tandler (1902) described an epithelial proliferation in all hollow organs in the 30 to 60-day foetus; normally these proliferating layers of epithelium which almost completely fill the lumen at this stage vacuolate later, re-establishing the lumen, and it has been suggested that a failure of vacuolation may lead to atresia. This would hardly explain those cases in which a segment is entirely absent, and it has been suggested rather that the pathological atresias of the mid-gut, like the normal physiological atresia of the vitello-intestinal duct which they closely resemble, may be due to the imposition of a longitudinal tension between mid-gut and yolk sac (Aird, 1939).

3. FREQUENCY

Atresia of the small intestine may be expected to occur once in approximately 20,000 births.

4. MORBID ANATOMY

Varieties

The atresia may take the form of a valve with a narrow lumen, of a diaphragm or of a reduction of the lumen to a microscopically narrow tube. Sometimes the bowel is replaced by a fine solid, fibrous cord, or a segment may be entirely absent, the bowel ending as a blind pouch above the deficiency and recommencing as a narrow tube below it. Usually, only one segment suffers, but sometimes several are affected. The jejunum is the seat of atresia more commonly than the ileum.

Pathological effect

The bowel above the affected segment dilates greatly; the bowel below remains empty with a narrow lumen not readily distensible.

5. CLINICAL PICTURE

Atresia produces the signs of obstruction soon after birth. If the upper jejunum is affected vomiting predominates and distension is slight; if the lower

ileum suffers, distension is early and vomiting late. Meconium is usually passed once soon after birth, and not thereafter. In later stages the signs of dehydration appear.

6. SPECIAL AIDS TO DIAGNOSIS

Examination of meconium

Ladd (1933) has shown that if meconium is passed after birth, useful information may be obtained from it. Normal meconium contains mucus, bile, keratinized epithelial squames, lanugo, hair and vernix caseosa. If a smear of meconium, stained for one minute with Sterling's gentian violet, fails to show stained squamous cells, atresia may be presumed.

7. DIFFERENTIAL DIAGNOSIS

Congenital pyloric stenosis

Volvulus neonatorum

The presence of bile in the vomit no less than the onset of obstructive signs as early as 1–3 days after birth serve to distinguish the condition from congenital pyloric stenosis. The signs of upper jejunal atresia are indistinguishable from those of neonatal volvulus except by examination of the meconium. Keratinized epithelial cells are present in the meconium of congenital volvulus.

8. TREATMENT

The treatment is operative, but operation should be preceded by the intravenous administration of water, salt and protein.

Technique of anastomosis

At operation, the lumen must be re-established by lateral anastomosis of the dilated bowel above to the narrow bowel below. There is a pronounced disparity between the diameters of the two segments, and the lumen of the distal segment may be so restricted that there is a risk of its occlusion by distortion at the suture line. To some extent distortion can be avoided by limiting the sutures to a single row of interrupted silk; it has been suggested also (Clogg, 1904) that a catheter, introduced by stab incision below the level of the proposed anastomosis and passed upwards to dilate the lower segment, be left in place while the anastomosis is performed, and withdrawn only on its completion.

Endotherm division of diaphragm

If at operation a diaphragmatic form of occlusion is found, it may be destroyed by an endotherm needle introduced through a longitudinal incision in the proximal dilated bowel just above the obstruction (Jones and Morton, 1938).

[Po]st-operative [tre]atment

After operation, nasal suction drainage should be established and saline infusion continued.

9. RESULTS OF TREATMENT

Congenital atresia is very often fatal. Davis and Poynter in 1922 collected 392 cases from the literature; all were fatal. Birgfeld in 1928 found 2 recoveries in a further 194 cases. Ladd in 1933 recorded 40 atresias of duodenum, jejunum and ileum, with 8 recoveries. Ileal obstruction is more likely to be fatal than is jejunal obstruction (Ladd and Gross, 1934).

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INTUSSUSCEPTION

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1. DEFINITION

200.] Intussusception consists of the passing of one portion of the intestine into another. It occurs in children or in adults, and the process may be acute or chronic.

2. AETIOLOGY

Causation

Intussusception in the adult is usually due to the presence of a neoplasm in the wall of the intestine protruding into the lumen and acting as a foreign body, but this group accounts for only 4 5 per cent of cases. About 80 per cent of cases occur under 2 years of age, and 70 per cent in the first year of life. The common varieties of intussusception are termed ileo-caecal, ileo-colic, enteric and colic. Under 1 year of age ileo-caecal and ileo-colic varieties form 99 per cent of all types of intussusception. The peak period for intussusception lie between the fifth and ninth month of life, so the causal factor must be due to disturbance of the normal anatomy, or to a change in the physiology of th child at that time.

ommon rieties

Anatomy

In the child under 1 year of age, the ileo-caecal valve protrudes into the caecum for ⅜ inch, and there is a ring of lymphoid tissue present in its sub-mucous coat The lumen of the caecum is not much larger than the lumen of the ileum at birth, and it bears no comparison with that of the adult, so that if there is any swelling of the lymphoid tissue increasing the protrusion of the ileo-caecal valve, it will readily act as a foreign body, and be gripped by the wall of the adjacent colon. In addition, the terminal inch of the mucosa

of the ileum is studded with masses of lymphoid tissue, which form a complete ring around the lumen of the gut. It is a common finding on reduction of the intussusception to feel the terminal inch of the ileum as a solid mass with no palpable evidence of a lumen. As the child enters its third year of life, there is a rapid subsidence of the lymphoid tissue, and the ileo-caecal valve no longer protrudes into the caecum. In ileo-caecal and in ileo-colic intussusception it is common to find a mesentery present in relation to the caecum and ascending colon. The excessive mobility given to the large bowel in this region would assist in the development of intussusception.

Mesentery

The incidence of intussusception is highest from the fifth to the ninth month of life, a period during which teething takes place, associated with gastro-intestinal upset, and in many cases, maternal milk has to be supplemented by patent milk foods to satisfy the growing needs of the child. The causation of ileo-caecal and ileo-colic types of intussusception is therefore due to the inflammatory swelling of the lymphoid tissue acting as a foreign body, so that peristalsis causes one portion of the bowel to enter into another. Owing to the distribution of the lymphoid tissue in the area, it is very difficult in many cases to distinguish between the two varieties.

Teething

The colic variety of intussusception occurs in older children up to about 7 years of age, and is due to the fact that the mucous membrane of the colon is present in excessive folds, studded with lymphoid tissue, and the lumen of the bowel is small enough to permit the inflamed swollen mucosal fold to be gripped when the muscular wall contracts. After 7 years of age the idiopathic type does not tend to occur, because the lymphoid tissue is diminished and the lumen increased. Cases of colic intussusception after this age are due to tumour formation in the bowel wall which acts as a foreign body. The enteric variety of intussusception is rare, and usually occurs in adults; it is due to prolapse of Meckel's diverticulum into the ileum, or to the presence of a tumour in the bowel wall. Haemorrhages into the bowel wall, as seen in Henoch's purpura, may act as a foreign body and may give rise to a rare form of enteric intussusception.

3. PATHOLOGY

Under 2 years of age, for practical purposes, we are dealing with the ileo-caecal and ileo-colic varieties of intussusception. The proximal portion of the bowel is received inside the distal portion of the bowel, with the result that there is an entering and returning layer called the intussusceptum, and an outer receiving layer called the intussuscipiens. The distal portion of the intussusceptum is called the apex, and the proximal portion of the intussuscipiens the neck of the intussusception (Fig. 45). In the ileo-caecal variety of intussusception the apex of the entering bowel will be the ileo-caecal valve, and the intussusceptum will grow at the expense of the invagination of the intussuscipiens. In the case of the ileo-colic intussusception the first portion of the intussusceptum is formed by invagination of the ileum until the pressure of the mesentery arrests it, and thereafter the intussusceptum grows by invagination of the colon (Fig. 46). This invagination process may take place to such an extent that in 28 per cent of cases the apex of an intussusception may be palpable on rectal examination.

Ileo-caecal and ileo-colic types

Invagination

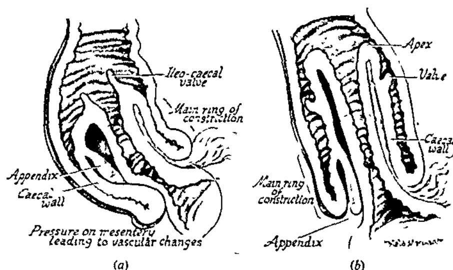


FIG 45 —(a) Ileo-caecal intussusception. (b) Ileo-colic intussusception. The ring of constriction is the neck of the intussusception.

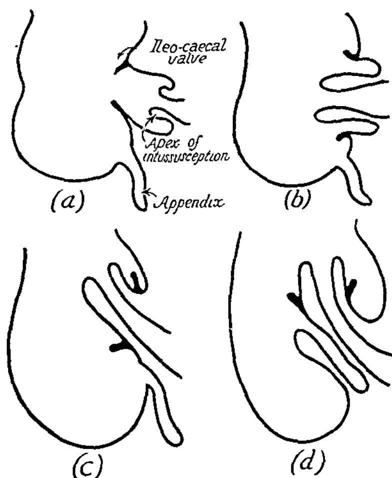


FIG 46 —Diagrammatic representation of the development of an ileo-colic intussusception.

As the ileum enters the colon the mesentery of the small intestine is dragged in and the vascular supply to the small bowel is upset by the pressure changes. Venous congestion first takes place, followed by an outpouring of fluid into the neighbouring tissues with resultant oedema. The peritoneum of the entering and returning layers becomes oedematous and inflamed, and adhesions may form which make reduction impossible. As the pressure increases there is an outpouring of blood-stained fluid into the colon, and death of the intussusceptum may take place owing to the shutting off of the blood supply. The devitalized bowel wall permits the bacterial content to spread through and set up a localized peritonitis followed by a generalized peritonitis. If no surgical treatment is given, the intussusception in the majority of cases will undergo gangrene and perforation, followed by a fatal peritonitis. In a few cases the intussusception may undergo spontaneous reduction; in other cases the intussusceptum may slough, and be passed per rectum. The ileo-colic intussusception is much more liable to early strangulation than are the other types, because of the extreme pressure placed on the bowel passing through the narrow ileo-caecal valve. The tumour is sausage-shaped in appearance, since the root of the entering mesentery is fixed in the midline over the lumbar vertebrae. The symptoms of early shock are due to the pull on the entering mesentery of the small intestine, and the muscular action of the affected segment of bowel is upset by the disturbance of its innervation as well as of its blood supply. Since the colon and the ileum are involved in the invagination process, the appendix forms part of the intussusception and undergoes to a lesser degree the effects of the pressure changes on its blood supply.

Vascular supply

Adhesions

Gangrene and perforation

Symptoms of early shock

4. CLINICAL PICTURE

The patient is usually a healthy well-nourished baby, who is suddenly seized with abdominal pain. The mother states that the child screams with pain, draws its knees up and becomes very pale. After a few minutes the colour returns, and the child appears to fall asleep. These attacks recur about every fifteen minutes, until the pain appears to become continuous. The child may vomit when the attacks are definitely established, but this is not a constant feature. Associated with these attacks, tenesmus may be present. One normal motion is passed, and then it is followed by the passage of blood-stained mucus.

Abdominal pain

Tenesmus

In the early stages, the pallor of the child, associated with the attack of abdominal pain, is the outstanding feature; it is only in the late stages that signs of dehydration are present, giving the eyes a sunken appearance and producing an apathetic expression.

General signs

An abdominal tumour will be felt lying obliquely or transversely, and maybe to the left of the umbilicus. The resistance of the muscles of the abdominal wall may mask the presence of the tumour, but continued gentle palpation will reveal it as it hardens during a spasm of pain. The tumour is tender to touch and handling it will set up a new spasm of pain so that continuous gentle palpation is the secret of success in assessing the presence of the tumour. A sensation of emptiness in the right iliac fossa on palpation is a very rare sign, and can only be present when the colon has been invaginated far from the original site of the ileo-caecal angle.

Abdomen

Rectal examination

In 28 per cent of cases the apex of the tumour will be palpable on rectal examination. The characteristic sensation imparted to the finger is the presence of a firm knob of tissue, the finger being able to pass around the circumference between it and the bowel wall. The presence of blood-stained mucus on the examining finger without the presence of faeces is a valuable sign, for it is a common finding that the bowel *acts once normally* after the onset of pain, and then obstruction supervenes. By the time blood-stained mucus is present in the bowel vascular pressure changes have taken place, and the lumen of the bowel is obliterated.

Blood-stained mucus



FIG. 47.—Barium enema, showing the cupola effect at the head of the barium mass and the shell of barium lying between the intussusception and the intussuscipiens.

In colic intussusceptions in children, which tend to be chronic, the intussusception may be so loose that the obstructive element does not arise and there may be passage of faeces without blood.

In the adult

As the majority of cases in the adult are due to tumours in the bowel wall, they may be either enteric or colic in type. The enteric group shows evidence of intestinal obstruction at an early stage, and the diagnosis is often made on opening the abdomen to relieve it. In the colic group the symptoms may not be so acute, and the patient may complain of obscure attacks of abdominal pain, associated with loss of weight and the presence from time to time of an ill-defined tumour.

5. SPECIAL DIAGNOSIS

Contrast radiography

Typical observation

There is obstruction to the injection of a barium enema, and a tumour may be palpable at the site of the obstruction. There is a cupola effect at the head of the barium mass, and a thin cylindrical shell of barium around the intussusceptum and inside the intussuscipiens. After the evacuation of the enema, there may remain a cylindrical shell of barium surrounding and outlining the intussusceptum (Fig. 47) This form of special diagnosis is very rarely required, and should be used only in cases of doubt when the tumour cannot be palpated under general anaesthesia.

6. DIAGNOSIS

In a baby the history of an attack of severe intestinal colic, associated with the presence of a palpable abdominal tumour and the presence of blood-stained

mucus on rectal examination, is absolutely diagnostic of the condition. In certain cases the abdominal tumour may be present, but not palpable, for it can become hidden under the liver.

7. DIFFERENTIAL DIAGNOSIS

(1) Enteritis

The onset of symptoms is not so acute, and there is usually a history of digestive disturbance. By the time blood-stained mucus is present in the stool, dehydration is usually a marked feature, vomiting is generally associated with the diarrhoea, and faeces are mixed with the blood-stained mucus. The crucial point in the diagnosis is to make certain whether complete intestinal obstruction is present or not. In a case of enteritis, the presence of an undescended caecum associated with enlarged ileo-caecal glands may simulate the presence of an abdominal tumour.

Digestive disturbance

(2) Henoch's purpura

Henoch's purpura may give rise to an attack of intestinal colic associated with the presence of blood-stained mucus, and an abdominal tumour may be palpable. The patient, however, is usually 7–12 years of age, and petechial haemorrhages are present in the tissues around the joints of the extremities. The abdominal tumour is due to infiltration of the bowel wall with haemorrhages and oedema; therefore it is fixed, and the apex of an intussusception is not palpable on rectal examination.

Petechial haemorrhages

8. PROGNOSIS

Spontaneous reduction of the intussusception may occur in about 1 per cent of cases in children, but the majority of intussusceptions require surgical intervention in order to reduce them, and the operation carries a mortality rate of about 1 per cent if the duration of the symptoms is not longer than 24 hours. The time factor is of great importance, because the longer the intussusception has been present, the less likely is it to be reducible at the time of operation. As soon as the intussusception in a child becomes irreducible, the mortality rate associated with the treatment rises to 80 per cent. The ileo-colic type of intussusception is the most difficult to reduce, owing to the pressure changes occurring round the narrow ileo-caecal valve

Surgical intervention

Mortality rate

9. INDICATIONS FOR SURGICAL INTERVENTION

As soon as the diagnosis of intussusception has been made, surgical intervention is required. In my opinion conservative treatment by hydrostatic pressure has no place. Some authorities claim that 50 per cent of cases of intussusception can be reduced by this method. They state that the child should be under general anaesthesia, when the douche-can with a column of water 3½–4 feet high is used. The reduction is usually assisted by manipulation of the tumour through the abdominal wall, and in all cases preparation is made to carry out immediate operation if it is not certain that reduction has taken place As the pathological changes vary from case to case, and as the evidence for reduction by conservative measures must always be in doubt, I am convinced that operative procedure is indicated in all cases in which the diagnosis of intussusception has been made.

Hydrostatic pressure

Manipulation of tumour

10. PRE-OPERATIVE MANAGEMENT OF THE PATIENT

Intravenous infusion of saline

Operation should take place as soon as possible in order to prevent ischaemic changes in the gut. If [illegible] venous drip infusion [illegible] the operating theatre, [illegible] time should not be lost in trying to gain full correction of the fluid balance.

11. OPERATIVE TECHNIQUE

(1) Reducible intussusception

Anaesthetics

A general anaesthetic consisting of gas, oxygen and ether should be given. Spinal anaesthetics, in cases of intussusception in children, should not be given, as the patient already may be suffering from a severe degree of shock.

Incision

A low right paramedian incision is made, and on opening the abdomen, two fingers of the operator's hand are gently introduced, and no intestine is allowed to escape on to the abdominal wall. Palpation of the colon on the left side will soon reveal the apex of the intussusceptum.

Reduction of intussusception

The intussusception is reduced by milking the colon distal to the intussusception in a retrograde fashion, so that the apex retreats from the surgeon's fingers. It is advisable to carry out this procedure slowly because otherwise the apex will run ahead too fast and will be lost in the region of the splenic flexure. The manipulation should take place entirely within the abdomen until the last few inches of the intussusception are left in the ileo-caecal region. This portion is brought outside the abdomen, and the reduction is carried out under vision. It is at this point that the intussusception tends to become irreducible.

Pressure on apex

The reduction should be persisted in by pressure on the apex of the intussusceptum; the entering ileum should not be pulled upon. Reduction by pressure should be continued even though splitting of the peritoneal coat is taking place, for the application of Sulphamezathine powder and a few catgut sutures in the split peritoneum after reduction will suffice to cure the condition.

Oedema

Relief from oedema in the intussusception can be procured in some cases by wrapping the intussusception in a warm moist towel, and applying equal pressure with the palms of the hand.

Adhesions

At the same time in difficult cases a blunt dissector introduced into the neck of the intussusception may free a few of the newly formed adhesions between the entering and returning layers. A further method is to divide the neck of the intussusception with a pair of scissors at one point in its circumference; this may relieve pressure at the critical point. If no success is obtained a finger can be introduced into the hole thus made, and reduction completed by

Direct pressure

direct pressure on the intussusceptum. If these methods fail, whatever further method of treatment is carried out for the irreducible intussusception, a mortality rate of about 80 per cent must be faced.

(2) Irreducible intussusception

Gangrene

In infants and in children an irreducible intussusception has usually undergone gangrene in one or more portions. Many methods have been advised for dealing with this lesion. They may be briefly noted as follows.

(a) Resection with lateral or end-to-end anastomosis.

(b) Resection with double enterostomy (von Mikulicz, 1903; Paul, 1900; Hartmann, 1907).

(c) Resection of the intussusceptum through an incision in the intussuscipiens, with or without lateral anastomosis (Barker, 1892; Jessett, 1892; Maunsell, 1892; Coffey, 1907).

(d) Lateral anastomosis about the lesion with secondary resection.

(e) Ileostomy with secondary resection.

(f) Lateral anastomosis about the lesion with secondary sloughing or healing (Rutherford, 1909; Parry, 1908–09; Montgomery and Mussil, 1930).

High mortality

High mortality is due to the fact that the irreducible intussusception is usually the late case, with potential gangrene of a mass of tissue in the peritoneal cavity. The child is suffering from shock, dehydration and toxaemia and the operative technique required to deal with the condition turns the operation into a major procedure. The intussusception should be removed since it is the source of the toxins. Direct anastomosis of the ileum to the transverse colon leaves no safety-valve for the intestinal obstruction that may develop in the post-operative period due to paralytic ileus. In all cases in which resection and enterostomy is carried out, the loss of fluid from the ileal opening is so great that it leads to an electrolyte imbalance, and the child dies from inanition. Resection of the intussusceptum through the incision in the intussuscipiens is not a wise procedure, for by the time the intussusception has become irreducible there is devitalization not only of the intussusceptum, but also of the intussuscipiens.

Paralytic ileus

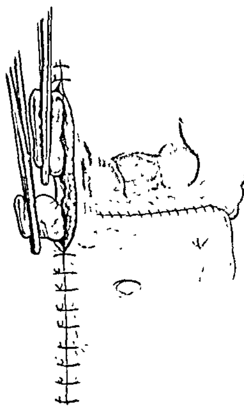


FIG 48.—Woodhall's modified double enterostomy operation for treatment of irreducible intussusception Intraperitoneal lateral anastomosis of ileum to transverse colon completed after resection of intussusception The clamped ends of the ileum and the transverse colon are brought to the surface and the abdominal incision is closed.

Advantages of Woodhall's operation

(g) Woodhall's modified double enterostomy operation offers the following advantages: (i) rapidity of execution, (ii) complete removal of the irreducible or gangrenous bowel, (iii) control of the concomitant intestinal obstruction, (iv) control of the loss of fluid and (v) restoration of the continuity of the intestinal canal (Woodhall, 1938).

Procedure

(i) Resection of the intussusception.

(ii) Lateral anastomosis of the ileum to the transverse colon. Anastomosis to take place 2 inches away from the divided ends of the transverse colon and ileum (Fig. 48).

(iii) The divided ends to be brought through the upper end of the abdominal incision and the lower part of the abdominal incision to be sutured separately. Clamps are maintained on the two ends of the bowel, and if distension arises, the clamp on the ileum can be released for a short period. In 6 or 7 days, when the intra-peritoneal anastomosis is functioning well, the ends of the bowel can be closed and replaced extra-peritoneally.

12. POST-OPERATIVE CARE

Correction of fluid balance

If the child has suffered from dehydration prior to the operation, the intravenous drip infusion of saline should be continued until the fluid balance is corrected. As the bowel has undergone a certain degree of inflammation, feeding should be restored gradually over a period of 4 days, feeds of glucose water only being given in the first 24 hours. In some children, after reduction of the intussusception, hyperpyrexia up to 105 degrees F. is seen; this appears to be due to absorption of toxins from the released intussuscepted area.

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[References to other titles are given under Intussusception in the Index Volume. The subject of Intussusception is also dealt with under the headings of Acute Intussusception *and* Intestinal Obstruction in the *British Encyclopaedia of Medical Practice* (1936), Vol. 1, p. 35 and (1938), Vol. 7, p. 230.

ISCHAEMIA

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1. DEFINITION

201.] Ischaemia is a condition in which the arterial blood supply to the tissues is reduced to a point below that necessary for their full function and nutrition. This interference may be just sufficient to give rise to symptoms, or may vary in extent up to complete obstruction.

2. AETIOLOGY

The causes, which are many and varied, may be classified as: (1) pressure from without; (2) pressure from within the tissues; (3) causes in the walls of the vessels; (4) causes in the lining of the vessels; (5) causes in the lumen.

(1) Pressure from without

Localized ischaemia due to direct pressure is frequently seen in areas of the body supporting the patient's own weight, more especially where bony prominences are immediately subjacent to the skin. Prolonged immobilization is sufficient, but rapid onset can be expected when the general health is poor or when neurological or vascular lesions are present. Similarly, the unrelieved pressure of a hard splint, a plaster-of-Paris cast or even a leather shoe, will produce symptoms in a few hours. *Decubitus*

Generalized and extensive ischaemia follows the use of any form of constriction when applied to a limb too tightly and for too long a period. The tourniquet has long been recognized as dangerous in this respect but equally so are ill-applied bandages. The bandage is usually satisfactory at the time of application but movement of the limb afterwards, for example at the elbow, may so alter the local conditions that the bandage becomes a tight and effective tourniquet. The well-applied bandage, especially when used for haemostasis, may act in the same way if appreciable traumatic oedema or inflammatory swelling occurs and so may an insufficiently padded splint or a plaster cast which completely encircles the limb. *Constrictions*

Strangulation In the abdomen, constriction and torsion of vascular pedicles produce at first an obstruction to the venous return, with engorgement and oedema of the affected viscera. The consequent increase in tension eventually strangulates the arterial supply and unless the condition is relieved within a few hours thrombosis occurs in the vessels and gangrene is inevitable.

Ligation Ligation of any artery renders ischaemia possible, and the incidence and degree of severity depend upon the site of the ligature and the health of the collateral circulation.

(2) Pressure from within the tissues

Tumours Localized ischaemia due to internal pressure is seen at the point of rupture of any abscess, and in a more chronic form in the tissues overlying steadily growing tumours such as exostoses and malignant disease.

The deep fascia is sufficiently dense in many parts of the body to act in the same way as a plaster cast when swelling occurs beneath it. In the popliteal *Extravasation* fossa an extravasation of blood or inflammatory exudate can so compress the vessels that ischaemia up to the point of gangrene may occur.

(3) Causes in the walls of the vessels

Spasm Spasm of the musculature of the blood-vessels may be intermittent, causing symptoms after many years or it may be persistent, giving an acute onset of ischaemia.

Intermittent spasm produces the Raynaud phenomenon, which may have no ascertainable cause or may be due to atherosclerosis, thrombo-angiitis *Cervical rib* obliterans or the effect of high-speed vibrating tools A cervical rib lifting up the subclavian artery may give rise to thrombosis in the artery in about 5 per cent of the cases which come to operation, but more often the vascular symptoms are due to spasm caused by irritation of the sympathetic nerve supply running in the lower trunk of the brachial plexus.

Cryopathy Cold normally produces constriction of the blood-vessels but minor degrees of cooling will produce spasm in sensitive individuals giving, when the arterioles are affected, the symptoms of perniosis and, in the case of the smaller arteries, the Raynaud phenomenon. Areas of the body with much fat and scanty muscular tissue have a poor blood supply, and the effect of cold is to cause saponification of the fat (fat necrosis). This occurs in the fat leg of a young woman with erythrocyanosis, and in the limb paralysed by anterior poliomyelitis. Extreme cold will cause such complete and prolonged spasm that damage to normal tissues is inevitable, giving the clinical picture of frost-bite, and a similar effect with similar symptoms is produced in ergot poisoning.

Trauma Slight localized injury may be irritative in effect and widespread persistent spasm will appear in the peripheral circulation. A supracondylar fracture of the humerus may cause no more than a small intramural haematoma in the brachial artery, but its presence will be indicated by absence of pulsation in the arteries of the forearm, together with widespread ischaemia. Similarly, a high-velocity shell fragment may not actually wound an artery but its concussive effect may determine an intractable and persistent spasm of the vessel walls. Gross trauma will either sever the artery or so damage its walls that thrombosis determines the amount of subsequent ischaemia.

(4) Causes in the lining of the vessels

Thickening of the intima is most frequently due to atheroma, in which syphilis plays a decreasing part, but in the younger patient thrombo-angiitis obliterans is the usual cause. Any thickening encroaches on the lumen of the vessels, but it is when ulceration occurs and a white thrombus is laid down that serious obstruction begins. The blood flow becomes so sluggish that complete obstruction by a red thrombus is probable, and the sudden onset of severe symptoms even to the point of massive gangrene will follow a long period of gradually increasing ischaemia in the limb.

Atheroma and thrombo-angiitis obliterans

(5) Causes in the lumen

The causes of blockage of the vessels which are derived from the lumen take the form of emboli. One variety is the clot from the wall of the left auricle or ventricle in mitral disease and this is usually set free when, as a result of treatment, the heart beat becomes more regular and powerful. Emboli also arise from atheromatous ulcers in the larger arteries, particularly the aorta, and multiple clots may be swept off by the blood stream.

Embolism

3 PATHOLOGY

The effect of interfering with the blood supply to the tissues is the same irrespective of the cause, but varies according to the extent of the interference. Complete stoppage means death of the tissues, but with less than this, survival is possible though in altered form.

(1) Death of tissue

Death of tissue, or gangrene, is clinically divided into dry and moist varieties but there is no essential difference It is largely a matter of the presence or absence of infection and the amount of fluid in the tissues. With an acute onset the capillary bed and the venous side of the circulation are left engorged with fluid, giving a moist gangrene, but in a long-continued and gradually progressive lesion such as atherosclerosis there is a tendency to form dry gangrene.

Gangrene

(2) Survival in altered form

When the survival of tissue is possible the essential change is a sacrifice of more highly specialized tissues such as secretory glands, muscle fibres and nerve endings. Nerve cells are particularly susceptible and will not recover if deprived of blood for more than a few minutes. Nerve fibres are more resistant but some peripheral nerves, such as the ulnar and the external popliteal, are more likely to suffer than other nerves in the limb Paralysis of these nerves may be the only evidence of nerve damage when the main artery of the limb is ligated, and following the prolonged application of a tourniquet, but when the latter is applied with excessive force the additional trauma of direct injury to the nerve fibres will complicate recovery. The supporting fibres need little blood, and they survive, but the specialized tissues are replaced by fibrous tissue and the whole area becomes sclerosed. Clinically this process is seen in the sclerodactyly following severe and repeated spasm of the digital arteries, in the sclerosis which follows the too-tight plaster case, and in the fibrosis following frost-bite. The skin becomes thin and glossy, losing its

Nerve palsy

Scleroderma

wrinkles, and is tightly stretched over joints and bony prominences where it is prone to ulceration. The digits are tapered through atrophy of the pulps and fixed in flexion by contraction of the fibrous tissue. The nails cease to grow and become opaque and discoloured with heaping-up of the nail-bed In the face, the skin is tightly drawn over the nose and the opening of the mouth progressively diminishes. The bones show osteoporosis, which may be severe, but in a few cases with severe Raynaud phenomena there is a deposition of calcium in the subcutaneous tissues especially in the fingers. With the increase in fibrous tissue, movement at the joints is restricted, with final fixation in a contracted position.

Calcinosis

4. CLINICAL PICTURE

Acute onset

The signs and symptoms vary with the acuteness of the onset of ischaemia, and also with its completeness and extent. With an acute onset, whether due to spasm or to occlusion of an artery, the initial sign is a pallor of the skin associated with a subjective sensation of coldness. Pain is absent at the beginning but there is progressive loss of sensation. With complete cessation of the blood flow a marbling of the skin occurs in 5–6 hours merging into a persistent cyanosis with superficial blisters. Pain commences and is of an intense burning character, accentuated by warmth. If muscular tissue is involved pain will appear earlier owing to the accumulation of metabolites in the muscles, and the more the muscles are used the more rapidly will pain appear. This pain is severe and cramp-like and rapidly becomes unbearable.

Complete obstruction

Incomplete obstruction

With incomplete stoppage coldness and pallor of the skin occur, but the cyanosis may not be severe. Pain is variable and when the muscles have been immobilized by splinting or a plaster-of-Paris case it may be ascribed to other causes, with the result that it is only when the fibrosis becomes manifest that the presence of ischaemia is suspected. The detection of the absence or diminution of the arterial pulsation in the affected limb will indicate the true cause of the pain.

Chronic onset

Pallor

When the onset of ischaemia is gradual the signs and symptoms arise so insidiously that usually it is pain which makes the patient seek advice. The limb will be much colder than its fellow and will show pallor on exertion, if not at rest. This pallor on exertion is not due to spasm of the vessels but signifies an arterial supply so diminished that it is unable to compensate for the removal of blood from the veins by muscular action. With further deterioration rubor appears in the dependent limb especially in the toes. Rubor, the peculiar brick-red colour of the skin, does not appear until a high degree of arterial obstruction is present, sufficiently severe to cause a coldness of the extremity such that the oxyhaemoglobin cannot dissociate. Consequently the capillaries and venules convey bright red blood, and since the oxygen is [illegible]

Rubor

Cyanosis

[illegible] than in the depth of colour Persistent cyanosis commencing in the extremity of a limb and not responding to warmth is a serious sign. Later the colour is even deeper, verging on black, the blood cannot be expressed from the capillaries, and gangrene is imminent.

There are three types of pain peculiar to ischaemia—intermittent claudication pain, rest pain and erythralgia.

Intermittent claudication may affect any muscle group but the muscles of the leg are most frequently involved. The pain is due to the stimulation of nerve endings in the muscles by metabolites which accumulate during exercise owing to the inadequate blood supply. A few minutes' rest allows the blood stream to wash away these factors and lower their concentration below the pain-producing threshold. The longer the rest period required the more inadequate is the blood supply, and deterioration is indicated by a shortening of the walking distance, and by the pain appearing in the thigh and proximal muscles.

Intermittent claudication pain

Rest pain is a late and ominous symptom. It is most commonly situated in the toes, especially the hallux, and does not disappear with rest. It is most troublesome at night and is due to slow and extreme tissue starvation.

Rest pain

Erythralgia is a reactionary pain which, therefore, does not occur in a progressive ischaemia. It is always described as burning and intense and is aggravated by warmth and dependency. It is the pain which is so troublesome to the patient recovering from an attack of the Raynaud phenomenon or frost-bite and which prevents the bringing of an ischaemic limb to the warmth of a fire.

Erythralgia

With ischaemia various nutritional changes arise in the tissues. The decrease in blood supply produces atrophy and fibrillary tremors in the muscles. Ulcers appear around the misshapen nails and on the atrophied tips of the fingers. The ulcers are extremely painful and show no tendency to heal. In the feet the most important change is indolent ulceration or paronychia of a toe, usually the hallux. More extensive necrosis takes the form of gangrene of digits and portions of limbs. In the obstruction of large vessels gangrene occurs to a greater extent and further proximally than would appear from the skin necrosis, but with injury to superficial vessels as in the tight plaster cast, or with frost-bite, extensive skin destruction may be the main loss although fibrosis of the deeper structures will occur.

Nutritional changes

Gangrene

5. SPECIAL AIDS TO DIAGNOSIS

A soft-tissue skiagram of a limb will show calcification of the arteries and differentiate between atherosclerosis and thrombo-angiitis obliterans, but if a radio-opaque medium, such as Pyelosil, is injected into the main vessel the actual site of the obstruction can be visualized. A skiagram will also show the calcium deposits in the pulps of the fingers in calcinosis (hypodermolithiasis). Skin temperature estimations by thermocouple are useful but the hand of the observer is sufficiently accurate for diagnostic purposes. A deficiency of the blood supply to a muscle group can be demonstrated by working the muscles on a simple ergometer and producing the pain of intermittent claudication. The oscillometers of the Pachon type are disappointing and investigations into the rate of blood flow through a limb are now being made, using tracer elements and detecting their presence by the Geiger counter.

Skiagrams

Skin temperature

Ergometer

Tracer elements

When treatment is being considered, an injection of Novocain into the sympathetic supply of the limb will show the maximal dilatation which can be obtained by section of these nerves, and this dilatation can be estimated by the plethysmograph or by the rise in skin temperature. In the lower limbs dilatation can be obtained very simply by the administration of a spinal anaesthetic.

Sympathetic block

6. TREATMENT

The essential in all successful treatment is the removal of the cause as soon as possible. This implies the recognition that ischaemia is present, and delay in this recognition is a major difficulty in initiating treatment.

Crush syndrome

Constrictions, such as those produced by tourniquets, should be released by the end of an hour and, although this time can be exceeded, if much tissue is involved the possibility of the development of a crush syndrome after release of the tourniquet must be borne in mind. We have known severe symptoms of this syndrome follow the release of a thigh tourniquet after 3½ hours and death occurred when, inadvertently, a similar tourniquet was left on for 11 hours. Direct pressure on the tissues requires relief in 4 or 5 hours, though, the weaker and more debilitated the patient, the shorter this time becomes. The unpadded plaster cast which encircles the limb is particularly dangerous and only careful watching, with splitting or removal of the plaster when there is any doubt, will avoid a catastrophe. Similarly, haematomas in the tissues require recognition and incision and evacuation of the clot if the condition is not completely satisfactory.

Emboli

Spasm

When the vessels themselves are at fault the cause usually cannot be removed, but an exception is the embolus, removal of which within 6 hours gives gratifying results. In the arm it is doubtful whether embolectomy is ever necessary because of the excellent collateral circulation, and the tendency is to adopt expectant treatment. Spasm of a large artery can defy all attempts at relief. If trauma has produced an intramural haematoma, a segment should be excised, but in the absence of a local cause the effect of a spinal anaesthetic or blocking of the relevant sympathetic ganglia with Novocain must be tried. If ineffective, the spasm may finally be broken by forcible distension of the artery with normal saline, but occasionally the condition fails to respond to any form of treatment.

Nursing

The nursing of the ischaemic limb is important and the not uncommon practice of wrapping up the limb in Gamgee tissue is harmful. Warming of the tissues increases their metabolic rate and calls for more blood. If this is not available, pain is increased and a tissue which might otherwise survive dies. It is therefore imperative that the limb be kept cool, but active cooling must not be practised. Similarly, elevation is contra-indicated because, with a dependent posture, gravity assists the blood flow and pain is eased. When there is much oedema elevation to the extent of laying the limb flat on the bed has to be adopted, but pain will arise, or be increased, and strong sedation becomes necessary.

Sympathectomy

For the chronic occlusive diseases, Buerger's exercises and contrast baths of hot and cold water rarely help; the intermittent venous-occlusion machines or the passive vascular exercise ("pavaex") apparatus are more useful. Relief is obtained in a small percentage of cases and the same applies to the use of injections of Padutin, but it has proved difficult to decide which case will benefit from any particular form of treatment. Sympathectomy in the form of section of the thoracic cord below the third ganglion or removal of the lumbar chain holds a definite place in treatment. Intermittent claudication cannot be affected because the muscles require so much blood, but minor trophic disturbances of the skin can be healed, and amputation may be averted

or postponed. Relief of rest pain is dramatic and produces such an improvement in the patient's general condition that his deterioration is halted. Results of the operation in the upper extremities are not so favourable as in the lower extremities, but a large percentage of the patients will state that the operation has been well worth while though the clinical result may be far from satisfactory. When nutritional changes have progressed to the stage of fibrosis of the tissues or widespread ulceration, these operations are valueless and should not be performed.

[References to other titles are given under Ischaemia in the Index Volume.]

JAUNDICE

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1. GENERAL

202.] Jaundice or icterus results from increase in the blood content of bilirubin or related substances sufficient to cause visible discoloration of the integuments. The term latent jaundice is applied to a lesser degree of pigmentation not appreciable to the eye.

Jaundice is not a disease but a symptom of many disorders of widely differing character. Thus in the diagnosis it is not only necessary to recognize that jaundice is present but also to determine its cause.

2. PHYSIOLOGICAL CONSIDERATIONS

For the present purpose it is sufficient to work on the basis of a simple version of the somewhat complicated physiological processes which enter into the mechanism of jaundice. The bile pigments (bilirubin and its oxidation product biliverdin) are derived from the iron-free molecule of the haemoglobin set free from disintegrating red blood cells. The conversion of haematoidin into bilirubin may be regarded as taking place in two stages. The first stage is believed to be effected by the cells of the reticulo-endothelial system, notably in the spleen, the bone marrow and the Kupffer cells of the liver; the second is believed to be effected by the polygonal cells of the liver. Jaundice may be due to excessive production of pigment, to interference with its secretion in the liver or to interference with its excretion through the bile-ducts into the duodenum.

3. TYPES OF JAUNDICE

Thus we may recognize three main types of jaundice.

(1) Haemolytic jaundice, due to over-production of pigment as a result of excessive haemolysis. This type of jaundice may result from various

haemolytic agents, such as potassium chlorate, and from infection by
lytic organisms as in streptococcal septicaemia. It is seen also in ac
jaundice (*see* p. 181).

(2) Jaundice due to liver damage, which may result from a wide vari
toxic and infective disorders. In this group come the jaundice which may
in the course of acute infective diseases, such as pneumonia, the jaundic
to arsenic, bismuth and many chemical products used in industry, the j
dice of acute yellow atrophy, and finally the group of diseases which incl
catarrhal jaundice, epidemic (virus) jaundice and homologous se
jaundice.

(3) Jaundice due to obstruction of the extrahepatic bile passages by sto
carcinoma or other lesions.

4. DIFFERENTIAL DIAGNOSIS

There are over 50 distinct causes of jaundice, but fortunately in the grea
majority of cases the diagnosis is readily made on consideration of the clinica
history or on collateral evidence.

When the cause of the jaundice is not immediately obvious, the diagnosis
usually lies between three common types of disease, to which may be added a
number of rarities. The three common types are (1) gall-stones in the common
duct, (2) catarrhal or virus hepatitis and other forms of hepatitis, and (3)
malignant obstruction of the bile-ducts. This last type must be subdivided
according as the jaundice is due to carcinoma of the head of the pancreas
(which may be amenable to surgery) or to other forms of malignant disease
for which only palliative measures are available.

5. INVESTIGATION OF A CASE OF JAUNDICE

To reach a diagnosis a full investigation is necessary, including a record of
clinical history, the physical examination and various special tests.

(1) Clinical history

Much may be learnt from an accurate clinical history and often the diagnosis can be made on the history alone. Particular attention should be paid to the mode of onset of the jaundice and to the presence of other symptoms such as pain, nausea or vomiting.

(*a*) *In stone in the common duct*

In stone in the common duct as a rule the history alone makes the diagnosis clear. There is a long history of biliary dyspepsia perhaps with recurrent attacks of severe biliary pain, culminating in a particularly severe attack as the stone is forced from the gall-bladder into the common duct. The jaundice makes its appearance usually a day or two after such an attack. Characteristically the jaundice is moderate or mild in degree, and it varies in depth from week to week or even disappears completely for a time. Pain often recurs, though less severe than the original colic, and often the pain is more central and pierces through to the back. There may be slight pyrexia, and even rigors, and there is some impairment of general health and strength, and some loss of weight.

Mild degree of jaundice

It should be remembered, however, that whereas this is the typical picture it is by no means invariable. Sometimes jaundice is absent for long periods, as the stone lies free in the duct and almost symptomless. Sometimes on the other hand, if cholangitis develops as a complication there is more marked jaundice accompanied by rigors, a high remittent *temperature and evidence of severe* toxaemia. In such circumstances the diagnosis is to be made from hepatitis, from hepatic abscess or portal pyaemia or even from Weil's spirochaetosis. Finally, in rare cases a stone impacts tightly in the common duct without much pain, and gives rise to progressively deepening jaundice which may simulate the picture of malignant obstruction.

(*b*) *In hepatitis*

Abrupt onset

In hepatitis as a rule there is no history of previous abdominal symptoms. The onset is abrupt, with nausea, profound anorexia and sometimes vomiting and diarrhoea. The jaundice appears within a day or two of the onset and *deepens rapidly to reach a bright orange tint. It persists for a variable period,* usually for 3–4 weeks and then gradually fades. In such cases the diagnosis presents no difficulties. Occasionally, however, the prodromal symptoms are less acute and the onset is somewhat insidious. Such a picture, especially when seen in an elderly patient, will arouse the suspicion of malignant disease.

(*c*) *In malignant obstructive jaundice*

Insidious onset

In malignant obstructive jaundice typically the jaundice develops insidiously and deepens gradually and progressively. Often the patient is unaware of the presence of icterus until his friends remark on it. As a rule, no other symptoms mark the onset, though when the patient is questioned he will recall that for a few weeks or months he has been failing a *little, lacking energy, having no* appetite and perhaps losing a little weight. He may also have noted such particular symptoms as itching of the skin and yellow vision. Pain may occur, though it is not a typical feature of malignant obstructive jaundice. When present it usually takes the form of a feeling of constriction in the epigastrium and lower thorax, as though from distension of the liver within its capsule.

While typically the symptoms in malignant obstructive jaundice are as described above, in rare cases the history is atypical. This is especially true of growths in the region of the ampulla of Vater which, *by reason of their* operability, it is particularly important to recognize at an early stage. In such growths, owing to the partial necrosis of the tumour, the obstruction may undergo temporary relief, and in consequence the jaundice *fluctuates or* may disappear for a time. In such growths also infection from the ulcerated *surface of the* tumour may spread to the bile-duct and liver and give rise to cholangitis with intermittent melaena, and may lead to a suspicion of duodenal ulcer or carcinoma of the stomach or colon.

Examination by daylight

When the presence of jaundice *is suspected the patient should* be examined by daylight or by a daylight lamp, for under ordinary artificial light even a considerable degree of pigmentation may be missed. Subsequent examination should be made under the same illumination to avoid a false *impression of* [illegible] for pigments [illegible] fresh [illegible] rectal specimen of faeces obtained conveniently on the gloved [illegible] examination.

(2) Clinical examination

A full routine clinical examination should then be carried out, bearing in mind that jaundice may be caused by such diverse diseases as carcinoma of the breast, bronchial carcinoma and Hodgkin's disease. Digital examination of the rectum may reveal an unsuspected growth in that situation, or nodules on the pelvic floor secondary to a growth in the stomach or colon. In women the possibility of a malignant tumour of the ovary must be borne in mind.

On abdominal examination particular attention is paid to the liver and gall-bladder. Some enlargement of the liver is usual in nearly all types of jaundice but marked enlargement is especially characteristic of hepatitis. Occasionally it is possible to feel the bossy nodules of malignant deposits, but this is a late feature and should not be relied on in the diagnosis.

Palpation of the gall-bladder

Palpation of the gall-bladder is particularly important. In jaundice due to carcinoma of the pancreas the gall-bladder is almost always distended, whereas in all other forms of jaundice it is of normal size or shrunken. Consequently this sign is useful not only in the diagnosis but also as a guide to treatment, for a distended gall-bladder signifies a growth which may, in favourable circumstances, be amenable to surgery. It is true that there are exceptions to Courvoisier's law—notably in jaundice due to common-duct stone where a second stone impacted in the cystic duct may lead to distension of the gall-bladder—but in general a palpably distended gall-bladder may be assumed to indicate a growth in the head of the pancreas.

Peritoneoscopy

Unfortunately the distended gall-bladder is not always readily palpable, especially if it is obscured by a prominent Riedel's lobe of the liver, or if it swings on a mesentery, and sags backwards towards the right kidney pouch. Consequently, although Courvoisier's law is theoretically reliable its practical value is less certain. In doubtful cases the state of the gall-bladder may be determined by direct inspection by peritoneoscopy. (*See* Endoscopy—Peritoneoscopy. Vol. 3, p 432.)

6. SPECIAL INVESTIGATIONS

Bilirubin estimation

In jaundice of obscure origin the patient should be kept under observation in bed. A 4-hourly temperature record is kept, for occasional slight pyrexia may point to a stone in the common duct. The Wassermann reaction is tested as a routine. The urine and stools should be watched for bile pigments. Every few days the bilirubin content of the serum is estimated, as a check on the progress of the jaundice; a steady fall points to a resolving hepatitis, a fluctuating index points to a stone in the duct, a steady rise points to malignant obstruction.

(1) X-ray examination

This should be done as a routine. A skiagram of the chest will exclude growths in the lung, and will show the level of the right dome of the diaphragm, which may be raised by an enlarged liver. A straight skiagram of the abdomen may demonstrate gall-stones, though it must be remembered that the majority of stones cannot be visualized in this way. Cholecystography is useless in deep jaundice, for if no bile is being secreted no dye can reach the gall-bladder. In some cases a barium examination will be needed to exclude carcinoma of the stomach or colon. In carcinoma of the pancreas the barium

meal may outline a deformity of the duodenal loop, and a growth at the ampulla may show a filling defect in the second part of the duodenum.

Liver function tests are sometimes of value in the differential diagnosis between hepatitis and malignant obstruction. (*See* Biochemical Tests, Vol. 2, p. 71.) In particular cases other laboratory tests are called for, such as the tests for Weil's spirochaetosis and stool examinations for intestinal parasites.

(2) Aspiration biopsy of the liver

Lastly, consideration must be given to aspiration biopsy of the liver. This is done by a special long fine-bore cannula, which is inserted under local anaesthesia through the tenth intercostal space in the mid-axillary line, and thence deep into the liver, punching out a core of liver tissue for histological examination. This method requires a good deal of care in its performance and is not entirely free from the danger of haemorrhage, but it is capable of yielding useful information, particularly in the early diagnosis between hepatitis and malignant obstruction of the bile-ducts.

7. RARER CAUSES OF OBSTRUCTIVE JAUNDICE

Apart from gall-stones and malignant disease the extrahepatic bile-ducts may be obstructed by rarer lesions. Enlarged glands in the porta hepatis may occur in Hodgkin's disease, leukaemia and occasionally tuberculosis. In such cases the diagnosis is based upon collateral evidence. Post-operative stricture of the common duct can be diagnosed readily on the history. Cystic dilatation of the common duct is a very rare abnormality, characterized by recurring jaundice associated with a large cystic swelling deep under the liver. A stone in the pancreatic duct may be recognized by its characteristic linear shadow, transversely disposed, as seen in skiagrams. Chronic pancreatitis simulates malignant disease very closely and cannot readily be distinguished from it. Exceptionally, the common duct may be obstructed by foreign bodies or parasites which gain access from the duodenum.

8. TREATMENT

The effective treatment of jaundice is the treatment of the causal condition, and this is discussed in the appropriate chapters. Here we shall deal only with the treatment in relation to the state of jaundice itself.

Pruritus

Pruritus is the most annoying feature of jaundice from the patient's standpoint, and the one for which he demands treatment most urgently, but although some relief may be gained by the application of calamine or other soothing lotions, no treatment yet devised has proved really effective. Fortunately the pruritus usually does not persist long, and tends to pass off in 5 or 6 weeks or so, even though the jaundice remains and grows deeper.

Pre-operative treatment

In severe jaundice, and particularly when operation is to be carried out, special measures must be taken to mitigate the effects of liver damage. The administration of amino acids, which was recently introduced for this purpose, unfortunately has not proved so effective as was expected. At the present time the most useful line of treatment is the well-established one of administering sugar in order to replenish the glycogen reserve, which is greatly diminished in all diseases affecting the liver. If the patient can take food the sugar may be given as sucrose in orange or lemon drinks or in tea or other

beverages. In other circumstances glucose may be given intravenously. If glycosuria results, to ensure full utilization the glucose may be backed by appropriate doses of insulin.

Haemorrhage after operation

Haemorrhage was formerly a complication to be feared after operation in jaundiced subjects, particularly in deep jaundice of long duration. The haemorrhage generally developed a few days after operation, and took the form of an ooze of blood into the depths of the wound, with the formation of a haematoma. Less often there was bleeding from the nose or other mucous membranes. The haemorrhage is now known to result from a deficiency of prothrombin, which may be due partly to faulty absorption of vitamin K from the intestinal tract (in the absence of bile) and partly to faulty synthesis in the damaged liver. The deficiency may be estimated by Quick's prothrombin test, which consists essentially in measuring the clotting time of the blood after adding an excess of calcium and thrombokinase. In such circumstances the clotting time of normal blood is very short (10–30 seconds according to the precise conditions of the test) but if prothrombin is deficient the time is correspondingly lengthened. Quick's test finds its main application in patients under dicoumarol treatment for thrombosis. (*See* Coagulants and Anti-coagulants, Vol. 3, p. 76.) In jaundice it is rarely necessary, and indeed may be misleading unless done daily, for after operation in a jaundiced patient the prothrombin index may fall suddenly and haemorrhage may occur when least expected.

The administration of vitamin K is highly effective in preventing haemorrhage in all except the most severe cases of jaundice, and since it is entirely free from danger it should be given as a routine in all operation cases. The dose is 10 milligrams, to be given intramuscularly. The first injection should be given 12 hours before operation, and repeated once or twice daily during the subsequent week.

9. ACHOLURIC JAUNDICE

Here the jaundice is due to excessive production of bilirubin (or a related substance) as the result of excessive destruction of fragile red blood cells. The secretion and excretion of bile are unimpaired, consequently the stools retain their normal pigmentation. Since the liver is undamaged the untoward effects of other types of jaundice are absent.

Congenital

Acholuric jaundice may be congenital or acquired. In the congenital form the jaundice persists throughout life and is accompanied by hypochromic anaemia resulting from the excessive red-cell destruction. In some cases both jaundice and anaemia are slight and the patient enjoys fair health; in others the patient is under-developed and delicate, and presents all the customary symptoms of severe bloodlessness.

Acquired

In the acquired type the disease more commonly takes the severe form and from time to time presents acute exacerbations—haemoclastic crises—in which blood-cell destruction proceeds apace, with consequent intensification of both jaundice and anaemia. In either the congenital or the acquired type the clinical picture may be complicated by the formation of pigment gallstones, which may lie in the common duct and add an obstructive element to the jaundice. For further details regarding the nature of this disease the reader is referred to the *British Encyclopaedia of Medical Practice*.

The diagnosis of acholuric jaundice is based upon the following findings: absence of bile salts from the urine, normal pigmentation of the faeces, negative result from the van den Bergh direct test, abnormal fragility of the red blood cells and excess of reticulocytes.

The spleen in acholuric jaundice is usually of normal size or but little enlarged, and it presents a normal appearance on microscopical examination. Splenectomy, nevertheless, is a highly effective measure of treatment, and in the great majority of cases is followed rapidly by disappearance of the jaundice and return of the blood picture to normal.

[References to other titles are given under Jaundice in the Index Volume. The subject is also dealt with in the *British Encyclopaedia of Medical Practice* (1938), Vol. 7, p. 261.]

JOINTS—ARTHROGRAPHY

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1. GENERAL

203.] Although the diagnosis of most internal derangements of the knee joint is obvious clinically, there is always a proportion in which a lesion of the menisci is doubtful or there is some doubt as to whether the lesion is in the medial or lateral meniscus, or possibly in both.

It is in these cases that air arthrography is of greatest value. It should always be remembered that it is only an aid to diagnosis, and must be taken into consideration with the clinical picture. For this reason, it is of advantage that the clinical examination and interpretation of the skiagrams should be by the same person.

(1) Routine examination

With this in view, a satisfactory routine is to re-examine the case clinically and perform an examination under anaesthesia at the same time as doing an air arthrography.

The routine is as follows: The leg is given the standard pre-operative preparation for 24 or 48 hours. The patient is taken to the theatre where the knee is reviewed clinically. Anaesthesia is induced with Pentothal, and the knee is examined with the patient relaxed. The preparation towels are removed and air is injected into the joint until it is full, but not tense.

(2) Injection of air

Before injection, the air is filtered through cotton-wool. The exact quantity of air injected is of little importance, but the usual amount is about 100 cubic centimetres It is of some importance that the needle should be inserted accurately into the joint. If several unsuccessful attempts are made, a small haemarthrosis may result, and any fluid in the joint, in excess of that normally present, is a contra-indication to air arthrography. Aspiration of fluid is not sufficient to allow reliable skiagrams to be taken as the small residue may produce misleading shadows.

Preparation towels are re-applied so that should a lesion be found, operation can be proceeded with on the following day if desired. The patient is then transferred to the x-ray department.

2. TECHNIQUE OF RADIOGRAPHY

(1) Apparatus used

The apparatus used is shown in Fig. 49. It consists of a sinus cone 2½ inches in diameter and 16 inches long and a stand with a curved top containing a curved cassette 10 inches by 8 inches.

(2) Routine views

The routine views of the menisci are shown in Fig. 50. They are four of the medial (M1, M2, M3, M4), and three of the lateral (L1, L2, L3). They are all tangential. Exposures of 55 milliamps, 45 kilovolts and 0 4 seconds are given.



FIG 49.—Routine view being taken. The apparatus is shown. View M2 is being taken. Note the tibia being abducted on femur to widen the medial side of the joint. (*J Bone Jt Surg.*)

A crêpe bandage is applied firmly over the suprapatellar pouch to express the air into the part of the joint being examined.

The knee is then placed over the curved stand and the sinus cone is focused

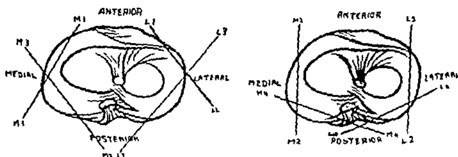


FIG. 50 —Routine views of menisci. All tangential. (*J. Bone Jt Surg.*)

on to the side of the joint being examined so that the central ray *will pass* through the joint.

The knee is slightly extended until the angle between the cone and the tibia is about 90 degrees. The side of the joint being examined is then widened by either abducting or adducting the tibia on the femur (Fig. 49).

The position of the joint is localized most satisfactorily by palpation. It is unnecessary to define it accurately by screening or marking the skin.

The usual view to take first is M2, that is, the antero-posterior view of the medial meniscus. The views M1 and M3 are obtained by rotating the leg through about 45 degrees to either side of the antero-posterior plane. The views L1, L2 and L3 are obtained similarly on the other side of the joint. View M4, however, is obtained by a different technique. For this, the patient



Fig. 51.—Technique for taking view M4. Note the knee at 90 degrees, the tibia being internally rotated on the femur.

is turned on to his side, the knee is flexed to 90 degrees and a flat cassette is placed beneath it. The tibia is then internally rotated on the femur, the sinus cone being focused on the posterior part of the joint (Fig. 51).

By internally rotating the tibia on the femur, the posterior part of the medial tibial condyle is shown up in outline with the meniscus lying on its posterior lip



Fig 52.—Method of taking "skyline" view of the patella with air between articular surfaces.

(3) View of patella

A further useful view may be obtained of the under-surface of the pa
In Fig. 52 is to be seen a dental occlusal film being pressed into the ligament.



(a)



(b)

FIG 53

The knee is flexed until the patella tap can only just be obtained. This may b assisted by placing the hands, covered with lead-impregnated gloves, o either side of the patella and pressing to raise it up from the femur. Rathe more air is required for this view than for the others, and even then it is no possible to obtain it in every knee.



(c)



(d)

FIG 53 —Routine views of a normal medial meniscus. (a) M1, (b) M2, (c) M3, and (d) M4. (*J. Bone Jt Surg.*)

3. INTERPRETATION OF SKIAGRAMS

(1) Normal appearance

The points to note are:

(a) The medial meniscus is attached at the periphery throughout, whereas the lateral one is attached at the periphery only at its anterior horn as in view L1.

(b) The lateral meniscus is very much larger than the medial.

(c) In view M4 the medial meniscus lies on the posterior lip of the medial condyle of the tibia, and the appearance suggests it is being drawn out of the joint slightly.

The actual skiagrams of normal menisci are shown in Figs. 53 (a)–(d) and 54 (a)–(c).



(c)

FIG. 54.—Routine views of normal lateral meniscus—(a) L1, (b) L2, and (c) L3. Note difference in shape compared with medial; also popliteal separation in views (b) and (c) (*J. Bone Jt Surg.*)

(2) Appearance of lesions

It is easier to diagnose lesions of the medial than of the lateral meniscus. This is due to the presence of the popliteal separation on the lateral side which must not be mistaken for a peripheral tear.

Bucket-handle tears

(1) Bucket-handle tears of either meniscus are seen without difficulty (Fig. 55). The outer rim is small and irregular and the displaced bucket handle is usually seen in the intercondylar notch. Shadows in this region may, however, be misleading, and it is safer, particularly on the medial side, to make the diagnosis more on the shape of the outer rim than on the presence of the

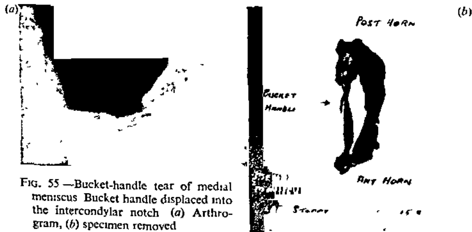


FIG. 55.—Bucket-handle tear of medial meniscus. Bucket handle displaced into the intercondylar notch (a) Arthrogram, (b) specimen removed

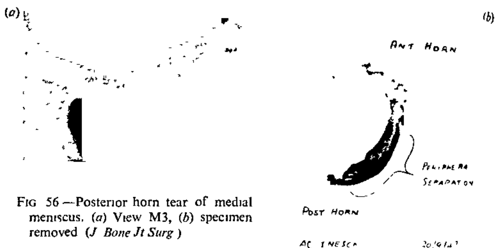


FIG 56—Posterior horn tear of medial meniscus. (a) View M3, (b) specimen removed (*J Bone Jt Surg*)



FIG. 57—Early split on under-surface only of medial meniscus. (a) View M3, (b) specimen removed



FIG 58.—Osteochondritis dissecans. Loose body separating from lateral femoral condyle.

displaced bucket handle. This is less important on the lateral side
the displaced portion is well seen and the outer rim less definite.
(2) Anterior and posterior horn splits are well seen in the appropriate v
(Figs. 56 and 57). On the lateral side, a peripheral tear consists of an elo
tion either forwards or backwards of the popliteal separation. In the art
gram this is shown as an abnormally wide separation with the menis
somewhat displaced towards the centre of the joint, the appearance suggest
that it would be trapped if the condyles were allowed to come together.
A variety of other lesions, such as ragged menisci, tags, erosion of articul
cartilage and loose bodies (Fig. 58), may also be seen. Occasionally, a disco
meniscus is found.

4. CONCLUSION

Air arthrography is a simple and safe procedure which can give considerable help towards obtaining an accurate diagnosis in cases of internal derangement of the knee joint, but it should be done only if rigid asepsis can be guaranteed.

[References to other titles are given under Joints—Arthrography in the Index Volume.]

JOINTS—CAISSON DISEASE OF

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1. DEFINITION

204.] This is a condition which should be borne in mind in dealing with men who have worked in caissons under compressed air—in digging tunnels, in underpinning buildings and so on; such workers are termed "divers" although the work may be entirely inland. As out-patients they may describe themselves as bricklayers and omit to mention the peculiar conditions of their work.

2. AETIOLOGY

Rules for decompression

Employers have rules for the decompression of these men, but rules are not always kept and attacks of "bends" are not uncommon. The men will be treated in the medical lock and will recover from the pain of bends. With bends, bubbles of gas (nitrogen) appear in the tissues and in the vessels. In caisson disease of joints, emboli, which block the arterial stream, occur in vessels towards the articular end of a bone and interfere with any anastomotic supply; thus infarcts of avascular necrosis are produced under the articular surface.

3. SURGICAL ANATOMY

Avascular necrosis

The blood supply of the femoral head is precarious and, therefore, any interference with it readily leads to avascular necrosis. The articular cartilage in a joint gets most of its nourishment from the synovial fluid.

4. PATHOLOGY

With deprivation of blood supply to a segment of the bone end, avascular necrosis may occur. If the part were kept at rest a fresh blood supply could develop, the necrotic area would be revascularized and no permanent damage need result. The whole process would be silent and devoid of symptoms. If the part is not rested the reaction of the surrounding bony tissues is sclerosis and [illegible] may
separation of th[illegible] may
be driven into [illegible] red,
result. The nourishment of the overlying articular cartilage is [illegible]

but with the giving way of the underlying bone it will be somewhat flattened, slightly yellowish in colour and rather wrinkled. Judging from the skiagram and microscopical section, it is not thinned or diminished in vitality.

Under the microscope the articular cartilage is not only well nourished but may be growing into the necrotic underlying bone, and in the section may give rise to a suggestion of sarcomatous change. The alteration in shape of the articular end will, in the course of months, give rise to changes of a degenerative type in the joint.

Degenerative changes

5. CLINICAL PICTURE

The typical history is that the patient was working in compressed air some 12 months or more previously and that he suffered from bends. After recovering from bends he may have returned to work, but symptoms have developed recently, either gradually or perhaps suddenly in one or more joints, especially the hip. There is nothing specific on clinical examination beyond the signs of arthritis in that joint, but the amount of reflex muscular wasting seems to be more than is commonly associated with the average degenerative joint lesion. The Wassermann reaction and tuberculin test are negative.



FIG 59.—The left hip of a man, aged 38 years, who had suffered from "bends" 4 years before this skiagram was taken. Note the wedge-shaped infarct in the femoral head, and decalcification with pseudo-cystic change in the neck Though there are degenerative changes in the hip, the joint space over the infarct is not diminished.

6. DIAGNOSIS

Apart from the history, a skiagram is essential. In the skiagram the end of the bone will show a dense patch, representing the original infarct from which the calcium salts have not been absorbed and which is shown up by the decalcification of the surrounding bone. This patch of avascular necrosis may be fragmented. It is probably driven into its bed and there is a zone of sclerosed bone beneath that bed. The joint space over this patch—as seen in the skiagram—will remain unaffected for years, but after 2 or 3 years the rest of the joint space will be diminished and there will be lipping at the margins of the joint as a result of osteoarthritic change (Fig. 59).

Osteoarthritic change

7. DIFFERENTIAL DIAGNOSIS

Other causes of avascular necrosis in the articular end of a bone are: (1) fracture, in which the history will be that of violence; (2) osteochondritis dissecans, which is rare in hip or shoulder: in this condition the affected portion is far more sharply defined, and there is usually a history of accident; (3) quite rarely some other form of "illness", without the history that the patient has worked in compressed air.

8. PROGNOSIS

Hitherto these cases have not come to the surgeon until the avascular patch has lost its chance of revascularization, the patch is loosened and the articular end of the bone is deformed. As time goes on the sequestrum seems to get smaller and more fragmented, but it is lying in a bed of fibrous tissue, and these changes are correspondingly slow. A period of 4 years makes comparatively little change in the sequestrum, as shown in the skiagram. On the other hand, if the sequestrum can be removed, then the degeneration in the joint slows up and one vizualizes the cavity being filled up by the activity of the articular cartilage.

9. INDICATIONS FOR SURGICAL INTERVENTION

Immobilization of joint

Prevention is better than cure. If these patches of avascular necrosis could be detected early and if the joint were then immobilized, the sequestrum could be revascularized and bone cells could spread into it without deformity occurring. If a hip or a shoulder were always examined radiologically 8 or 10 weeks after a local attack of bends, the condition could be detected and the joint could be immobilized in a plaster cast. Further, since some workmen are more susceptible to bends than others, susceptible men should not be employed in caisson work.

Alleviation of pain

Once the condition is established the treatment must depend upon the individual patient. If a sudden movement has dislodged the sequestrum in a shoulder, then removal of the affected portion and the overlying cartilage will be indicated. The treatment of a hip will depend upon whether or not the other hip is affected. The decision to recommend excision, arthrodesis or some other form of treatment will depend upon the condition of the lumbar spine and sacro-iliac joints. The removal of a loose body from the head of the femur may entail dislocation of the hip to get at it, a procedure which is severe and may produce further necrosis in that femoral head. It does seem, however, as if removal of a necrotic fragment alleviates pain because the joint is no longer subject to abnormal stress.

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JOINTS—INJURIES AND ACUTE INFECTIONS

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1. AETIOLOGY

Routes of infection

205.] A joint may be infected with pyogenic organisms in four ways.

(1) By an open wound of the joint, for example, a gunshot wound;

(2) By the infection of an open fracture which extends into the joint;

(3) By the spread into the joint of infection from an osteomyelitis of the neighbouring bone; and

(4) By direct haematogenous infection.

2. ANATOMY

Wounds: relative frequency in different joints

Any joint in the body may, in theory, be infected by any one of these four means. In practice, however, certain joints are much more prone to infection than others, either by direct or indirect wounding, or by organisms which

come from the neighbouring bone or from the blood stream. In a recent survey of 134 war wounds of major joints (Carney, Fitts and Kirby, 1946), the knee was by far the most commonly affected joint, with the elbow second in order of frequency.

DISTRIBUTION OF 134 INJURIES OF MAJOR JOINTS (CARNEY, FITTS AND KIRBY, 1946)

	NUMBER OF CASES	PERCENTAGE
Knee - - -	77	57·5
Elbow - -	21	15·7
Ankle - -	13	9·7
Shoulder - -	12	8·9
Hip - - -	11	8·2

As would be expected the superficial joints are more frequently wounded than those which are thickly covered with soft tissue. Wounds of joints are naturally more common in adults than in children.

Relative frequency of blood-borne infection

On the other hand, infection of joints from the neighbouring bone or from the blood stream is more common in children and adolescents. Infection occurs most often in those joints in which the capsule encloses a part of the metaphysis. Ellis (1946) states that it occurs between the ages of 4 and 14 years and that males are more commonly affected than females in the proportion of 3 to 1. The joints most commonly infected are those of the hip, knee, elbow and wrist, in that order. The hip is the joint which is by far the most frequently involved by "closed" suppurative or infective arthritis, since a large area of the femoral metaphysis is intra-articular. Haematogenous osteomyelitis most commonly attacks the metaphysis and spread from here to the joint is easy in the hip.

3. PATHOLOGY

(1) Bacteriology

Types of infecting organisms

When a joint is infected through a wound or a compound fracture, the nature of the infection varies with the circumstances of the injury. In this type of wound, however, as in other accidental wounds, the most dangerous infecting organisms are staphylococci and streptococci.

Blood-borne infections

When the infection is blood-borne, either directly to the joint or as an extension from the neighbouring bone, the pyogenic cocci are the organisms most often responsible. Staphylococci account for well over 50 per cent of the infections, streptococci for about 25 per cent, and the remainder are due to pneumococci and mixed infections (Ellis, 1946). These last may occur, for example, as a complication of otitis media in childhood.

Gonococcal arthritis

Arthritis as a metastatic complication of gonorrhoea is becoming increasingly less common as the treatment of the primary condition becomes more efficient; this type of arthritis very rarely suppurates. According to Lees (1937) it used to occur in 2–5 per cent of all cases of gonococcal urethritis, and its onset was from the third week to the third month of the disease. Untreated urethritis should, therefore, be suspected in any joint infection for which there is no obvious cause. Gonococcal arthritis yields very readily to treatment with penicillin.

(2) Morbid anatomy

There are three degrees of reaction of joints to infection—serous, sero-fibrinous and suppurative. The degree of reaction depends upon the virulence of the infecting organism in relation to the body's defence mechanism. *Types of joint reaction*

(a) *Serous reaction*

The synovium is congested and inflamed and the joint is distended with clear, free fluid which clots after withdrawal from the joint, and becomes a transparent or slightly translucent jelly. Organisms are rarely found in the fluid either on direct smear or on culture. Indeed, it is doubtful whether there are any organisms in the joint cavity; the fluid is in the nature of a toxic reaction to their presence in the synovium. This kind of infection commonly subsides completely, leaving no permanent effect on the function of the joint, such as adhesions or capsular thickening, but if not treated it may progress to either of the two other types of arthritis. *Serous arthritis*

(b) *Sero-fibrinous reaction*

The inflammation of the synovial membrane is more intense and the synovium is covered with flakes of fibrin. The joint effusion is cloudy and cellular, contains floating fragments of fibrin and sets into a firm contracting clot. *Sero-fibrinous arthritis*

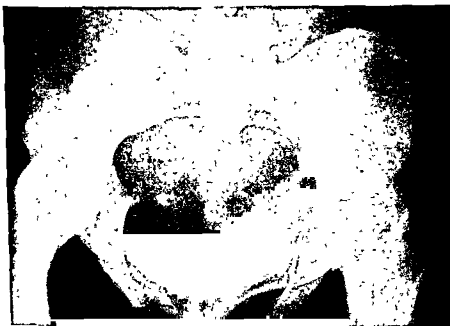


FIG 60.—Bilateral bony ankylosis of hips due to suppurative arthritis

Organisms can be found in the fluid, but these may be evident only in the direct smear. They may not grow on culture because of the natural bacteriostatic activity of the joint fluid (Florey and Jennings, 1944). Capsular thickening and joint adhesions readily result from this type of infection and frank suppuration may ensue.

(c) *Suppurative reaction*

With this serious form of reaction extensive damage may occur very rapidly in the joint. The synovial cavity is distended with pus of a high cellular *Suppurative arthritis*

content in which there are many organisms and much fibrin. The synovial membrane, the capsule and its ligaments and the intra-articular ligaments are all grossly inflamed and, as the tension of the pus within the joint rises, they may be wholly or partly destroyed. The pus spreads through the ruptured capsule and an abscess forms outside the joint. The articular cartilage is destroyed, and the cancellous bone underlying it is laid bare. Pathological dislocation of the joint is common, particularly of the hip joint in which the position of adduction and flexion, which is induced by the infective process, favours the occurrence of this complication. The epiphysis may be separated and die. Serious joint stiffness is likely to result from capsular fibrosis and intra-articular adhesions, and if much of the articular cartilage is destroyed the joint may ankylose by bone. It is imperative that this grave condition be brought under therapeutic control with all speed, and the possibility of ankylosis as an end-result should be constantly borne in mind (Fig. 60) Therefore the limb should be treated in the position in which it will be of most use to the patient should the joint stiffen.

Pathological dislocation

(3) Special types of arthritis

(a) Gonococcal

Gonococcal arthritis has been referred to already.

(b) Meningococcal

Meningococcal arthritis is rare and is similar to gonococcal arthritis; it usually yields readily to treatment with penicillin and never suppurates.

(c) Pneumococcal

Pneumococcal arthritis is a relatively rare condition, but presents a very definite clinical picture which was first noted in France (Plisson and Brousse, 1920) and was described by Feffer and Hirsh (1946). There is considerable peri-articular swelling, which is white in colour. There is marked distension of the collateral venous circulation and comparatively little reaction in the regional lymph glands. The joint fluid re-accumulates rapidly after aspiration. There is high fever, but otherwise the patient's general condition is much better than the severe local signs would lead one to expect. The condition yields readily to repeated aspiration and to intra-articular injection of penicillin.

(d) Typhoid

Typhoid bacilli sometimes produce a low-grade localized osteomyelitis. With rather greater frequency they cause an infective arthritis, usually of the hip joint. There is never suppuration, and the whole course of this complication of typhoid fever is mild—indeed it may pass unnoticed in the general illness. The damage to the articular cartilage, however, is usually considerable (this is seen in the marked diminution in the joint space radiologically) and secondary osteoarthritic changes soon develop (Fig. 61).

(e) Suppurative arthritis of infants

Suppurative arthritis of the hip in infancy pursues a different course from that occurring in adults or older children, mainly because the head and neck

of the femur are entirely cartilaginous up to about the seventh month of life. The infection is, therefore, not complicated by sequestration, but cartilage is very readily killed by suppuration, and extensive destruction of the upper end of the femur is common in this condition. The infection is blood-borne and the initial lesion is often in the skin or the umbilicus. The disease commonly is not associated with much pain and may escape notice in a febrile infant unless it is remembered and sought for. Diagnosis is established by the aspiration of pus from the joint. The infecting organisms are commonly the pyogenic cocci, and treatment by penicillin is indicated Penicillin should be



FIG. 61.—Left hip 7 months after arthritis occurring during typhoid fever Right hip clinically and radiologically normal.

given both parenterally and locally. Local administration of penicillin into the hip is not easy, and becomes more difficult as the joint swelling subsides, but the abscess cavity can usually be conveniently entered by a needle passed into the metaphysis of the femur through the cortex at the base of the greater trochanter (Aird, 1946). By this means penicillin can be given by continuous drip.

4. CLINICAL PICTURE

Clinical features

The clinical picture varies with the severity of the condition In its milder form, infective arthritis resembles acute rheumatism. The distinguishing feature is that only one joint, and that usually a large one, is involved. Acute rheumatism never affects only one hip. Sero-fibrinous and suppurative arthritis produce a much more definite and dramatic picture. The joint is swollen, hot and acutely tender. It is splinted by intense muscle spasm, and movement is exquisitely painful. The skin is reddened and there is superficial oedema. The regional lymph glands are enlarged and tender. There is high fever and the patient usually looks desperately ill. There is rapid muscular wasting, particularly of the affected limb. This is seen to its most marked degree in the quadriceps muscle when the knee is involved.

Early radiological appearances

Radiological changes are absent or slight in the early stages, and may never appear if the process stops short of suppuration with actual destruction of cartilage and bone. The only abnormal appearance seen in the skiagram in the early stages is osteoporosis, which is due to the inflammatory hyperaemia.

Late radiological appearances

When the joint suppurates and cartilage and bone are destroyed, marked radiological changes soon appear. The destruction of the cartilage is seen in the loss of the smooth, even contour of the joint surfaces; at this stage the joint space may not be narrowed even though the cartilage is destroyed, because the joint is held open by the fluid effusion. Death of the epiphysis is recognized by the fact that it does not participate in the process of decalcification which affects the surrounding bone; it thus appears relatively dense (Fig. 62). The bone underlying the articular surface is often destroyed and eroded, and as soon as the infection spreads outside the joint the characteristic appearances of osteomyelitis of the shaft of the bone appear. In particular there is subperiosteal formation of new bone, but when these

Epiphyseal necrosis



FIG. 62.—Suppurative arthritis of the hip in childhood showing epiphyseal separation.

radiological changes are seen, irreparable damage has been done. They *do not appear in the successfully treated patient.*

5. SPECIAL AIDS TO DIAGNOSIS

The white-cell count will help to differentiate various forms of acute arthritis. In suppurative arthritis there is always a marked leucocyte response. This, together with the clinical signs, may often make the diagnosis almost certain. Complete diagnosis, *however, must rest on an examination of the* aspirated fluid. This will show (1) the type of response in the joint, indicating the degree of virulence of the infection and (2) the nature of the infecting organisms. This information is of prime importance, because many of the organisms which commonly infect joints are highly sensitive to penicillin. Diagnostic aspiration of an infected joint should never be omitted.

Diagnostic aspiration

Sites of aspiration (Fig. 63)

(a) Shoulder

The needle is entered from the front, half an inch medial to, and half an inch below, the tip of the coracoid process. This landmark is readily *felt in*

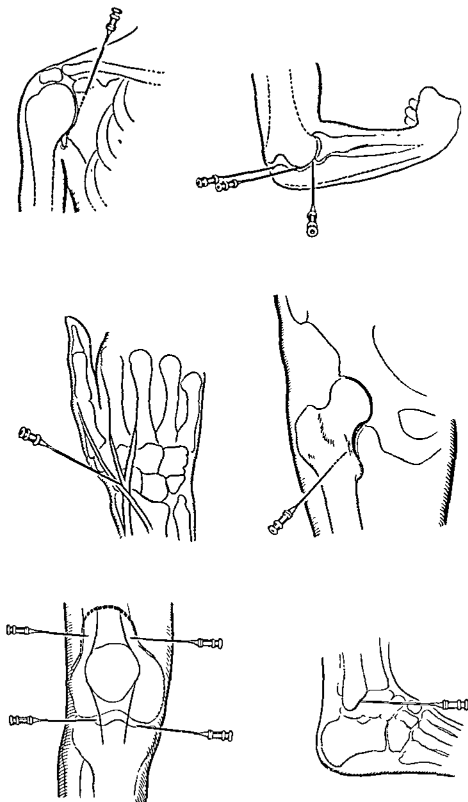


FIG. 63 —Diagram of sites of aspiration of joints. (*See* text.)

the delto-pectoral interval below the clavicle. The needle passes deeply with a lateral inclination of 30° and enters the lowest part of the shoulder joint where there is a loose pouch of synovium and capsule.

(*b*) *Elbow*

A needle can be passed into a distended elbow joint from behind, on either side of the olecranon, or from the postero-lateral aspect just above the head of the radius which serves as a guide. The latter is the more satisfactory site unless the joint is visibly distended.

(*c*) *Wrist*

The wrist joint is best entered by passing a needle from the dorsum between the tendons of the extensor pollicis longus and the radial extensors. This interval is easily felt, the radial styloid gives the line of the joint, and the needle passes forwards in an ulnar direction and inclined 45° in a proximal direction.

(*d*) *Hip*

The depth of the joint from the surface, the heavy covering of muscles, and the relatively small size of the peri-articular capsule make the hip joint difficult to enter with a needle, and unless the cavity is distended it may be impossible to find. The best place to attempt to enter the joint is the lower aspect where the joint cavity extends a variable distance along the neck. The needle is entered on the antero-lateral aspect of the upper thigh, opposite the lower edge of the base of the neck This point can best be found by palpation of the tip of the greater trochanter and reference to an antero-posterior skiagram of the hip. The needle passes directly inward and slightly upward, parallel with the lower aspect of the femoral neck.

(*e*) *Knee*

This joint presents no problem. It is easily entered from either side above the patella, or on either side of the patellar tendon.

(*f*) *Ankle*

The ankle is most readily entered by a needle passed directly back from the inner side of the front of the base of the lateral malleolus. Alternatively, a similar point on the inner side may be used, but the base of the medial malleolus is less readily palpated.

6. DIFFERENTIAL DIAGNOSIS

Importance of aspiration in differential diagnosis

Suppurative arthritis is a dramatic and almost unmistakable condition. It should be distinguished from para-articular osteomyelitis, but this distinction may not be possible as the conditions are commonly coexistent. The aspiration of pus from the joint establishes beyond doubt the diagnosis of suppurative arthritis.

When the arthritis is not yet frankly suppurative, several other conditions must be borne in mind.

(1) Haemarthrosis in haemophilia—usually the history of repeated haemorrhages and ecchymoses makes this diagnosis evident.

(2) Tuberculosis—this is usually a more indolent condition, with a longer history of trouble before advice is sought Often the radiological evidence of a destructive bone lesion leaves little doubt about the diagnosis. In the lower

limb biopsy of the regional lymph glands may establish the nature of the pathology (Arden and Scott, 1947).

Traumatic types of arthritis of hip

(3) Traumatic arthritis—the history is helpful, although the onset of inflammatory lesions of joints may be precipitated by an injury. Usually traumatic effusions subside very rapidly with simple rest in bed. The distinction may be particularly difficult in the hip joint, especially in children, in whom osteochondritis coxae (Legg-Perthes's disease) and slipping of the capital epiphysis of the femur may cause a painful limitation of hip movement. In traumatic arthritis there is usually no constitutional reaction, and no change in the number of white cells, or in the erythrocyte sedimentation rate.

Haemarthrosis

The single exception to this is when there is a large haemarthrosis, but the knee is the only joint in which the joint space is big enough to hold sufficient blood to produce general symptoms or signs, or alteration in the blood count or the erythrocyte sedimentation rate.

7. PROGNOSIS

Importance of early treatment

The sooner a joint infection subsides the better is the outlook for joint function. In connexion with wounds involving joints it has been stated that "if a patient can be given a cool dry joint twenty-one days after wounding, the prognosis should be that of a similar closed lesion" (Jones, 1945). The longer the infection lasts and the more intense the inflammatory reaction, the greater is the danger of limitation of joint movement by capsular and intra-articular fibrosis and adhesions. If much of the articular cartilage is destroyed, bony ankylosis may result. Epiphyseal destruction may cause deformity due to failure of normal growth.

8. TREATMENT

(1) General surgical principles

Treatment of wounds involving joints

When a joint is infected because of a wound or an open fracture which communicates with the joint, the first consideration is to treat that wound according to established surgical first principles—adequate surgical toilet and the provision of skin cover as soon as possible. The special points in primary wound surgery so far as the joints are concerned are: (a) the capsule should be closed by suture if at all possible, the wound in the overlying soft tissue and skin being left open if necessary; (b) in addition to whatever local and parenteral penicillin is given, 150,000 units of penicillin should be injected into the joint through a needle inserted from the sound side if possible.

Intra-articular penicillin

This injection should be repeated every 48 hours until it is evident that the danger of infection is past—usually after a period of about one week. If the wound is frankly infected, penicillin should be injected into the joint daily. If the fluid withdrawn from the joint indicates that the joint is infected it should be aspirated as often as is necessary to prevent distension of the joint (this will usually be not less than once a day) and 150,000 units of penicillin should be instilled into the joint after each aspiration.

(2) Parenteral penicillin

Parenteral penicillin

At the same time parenteral penicillin should be given in full doses. Although normal synovial membranes constitute a considerable barrier to the passage

of penicillin in either direction (penicillin instilled into a joint may persist in active concentration for 48 hours (Jones, 1945)), there is considerable evidence that penicillin passes readily either into, or out of, inflamed joints. Foster and Colquhoun (1945) showed that the fluid from inflamed joints contained a concentration of penicillin approximately equal to that of the patient's blood stream after intramuscular injection, and this has been confirmed by others (Bonnin, 1945; Hughes, 1945; Aird, 1946). Rammelkamp

Permeability of inflamed synovium to penicillin



FIG. 64.—Pathological dislocation in imperfectly treated suppurative arthritis of the hip. Note the subperiosteal new bone indicating extension of the infection along the diaphysis of the femur.

and Keefer (1943) demonstrated penicillin in therapeutic amounts in the blood stream after its injection into inflamed joints.

Haematogenous arthritis

When suppurative arthritis is blood-borne or is secondary to osteomyelitis in the neighbouring bone, the treatment is essentially the same. The joint should be repeatedly aspirated in order that it may not be unnecessarily damaged by the mechanical effects of high intra-articular tension; penicillin should be given both parenterally and into the joint; the joint should be adequately splinted. Because of the danger of stiffness, the joint should be splinted in the position in which the limb will be most functionally useful should movement not be recovered (*see* Arthritis—Surgical Considerations, Vol. 1, p. 393). In the hip, traction in a Thomas splint or, better, on a Jones abduction frame should be used because of the danger of pathological

Splinting

dislocation (Fig. 64). A plaster spica is not an adequate precaution against this disastrous complication.

(3) Open drainage

If suppuration in a joint does not subside in spite of repeated aspiration the joint should be drained by operation. This consists in incision down to the joint cavity and in opening the capsule. The capsule and the synovium should be left open, but a drainage tube must not be inserted into the joint. If any form of drainage is used it should be Vaseline gauze placed lightly in the wound, down to, but not into, the joint cavity. To insert a drain into a joint is to make subsequent stiffness of that joint almost inevitable.

Operative technique

(4) Surgical approaches

Shoulder.—The shoulder joint is best approached by an anterior incision in the delto-pectoral interval or through the anterior edge of the deltoid as described by Henry (1945).

Elbow.—The elbow joint is drained through a lateral incision over the head of the radius. Wide access can be obtained by detaching the lateral ligament from the lateral condyle of the humerus.

Wrist.—The wrist joint is opened from the back on either side of the extensor digitorum communis tendons.

Hip.—For reasons which have already been quoted, this is the most difficult joint to drain. The posterior approach has been advocated because theoretically it provides dependent drainage (Ober, 1924). It is doubtful whether this is of material importance. The muscles clothe the joint so completely that it is questionable if drainage is ever mechanically "dependent". The anterior approach is easier and less bloody, and the anterior wound can be made dependent by nursing the patient prone.

(1) Anterior approach—the lower two-thirds of the approach described by Smith-Petersen (1917) gives adequate access.

Smith-Petersen approach

(2) Posterior approach—an oblique incision is made over the back of the neck of the femur in the line of the fibres of the gluteus maximus and these fibres are separated. An areolar space is entered in which the sciatic nerve is found in the inner angle of the wound. The joint can be exposed by splitting the external rotators of the hip—the obturator internus, gemelli and the piriformis—in the line of their fibres. If more extensive exposure is needed these muscles can be detached from the back of the greater trochanter and mobilized inwards They then protect the sciatic nerve and the approach gives free access to the back of the hip joint.

Ober's posterior approach

(3) For severe and persistent suppurative arthritis of the hip, Girdlestone (1928) recommends excision of the head and neck of the femur through a lateral approach.

Excision of head of femur

Knee.—The knee joint is drained through a lateral incision above and below the patella. Occasionally posterior drainage may be needed.

Ankle.—The ankle joint is drained through an anterior incision between the peroneus tertius and the front of the lateral malleolus. The anterior talofibular ligament must also be divided in order to open the joint adequately.

(5) Post-operative care

Whatever other means is used in the treatment of an infected joint, rest of the joint is essential to recovery. Only rarely is rest in bed enough; the joint

Rest by splinting

must be splinted, and usually the most efficient method of splinting is an adequate plaster. The need for traction in hip-joint infections has already been noted. The splinting must continue until the joint is clinically and radiologically quiet, and then active movement is begun. The sooner this is begun the better is the ultimate outlook for a movable joint; but if movement causes a recurrence of the activity of the infection it must be discontinued. This period of resumption of active movement is a critical time, during which close supervision of the patient is necessary.

Active movement

Movement in fractures involving joints

In infected joints associated with open fractures, movement cannot always recommence as soon as the joint infection has subsided, but it should begin as soon as the fracture is clinically united. Radiological consolidation should not be awaited. During the period of immobilization the tone of the muscles of the immobilized limb must be maintained by regular active exercise; this must always be done when a limb is immobilized in plaster, and it aids greatly in the recovery of movement after the plaster is removed.

Active exercises in plaster

Position in which joints are to be immobilized

Because some degree of joint stiffness is common after suppurative arthritis, and because this may be of any degree up to bony ankylosis, it is essential that the limb be immobilized in the position which will be most useful should the joint become stiff. Sometimes a small remaining range of movement is painful and may cause recurring flares of the infection, and it may in these circumstances be better to arthrodese the joint by operation. This is one of the clear indications for the use of cancellous bone grafts instead of cortical grafts (Dick, 1946). There is no doubt that cancellous grafts are safer in the presence of actual or potential infection than cortical grafts (Abbott and his colleagues, 1947). Because of the greater ease with which they are revascularized they are much less likely to die and sequestrate.

Arthrodesis with cancellous grafts

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[References to other titles are given under Joints—Injuries and Acute Infections in the Index Volume. The subject is also dealt with under the headings of Joints, Diseases and Disorders in the *British Encyclopaedia of Medical Practice* (1938), Vol. 7, pp. 278 and 321.]

JOINTS—INTERNAL DERANGEMENTS OF THE KNEE

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1. INTRODUCTION

206.] The phrase, *internal derangement of the knee joint*, was coined by Hey [illegible]

picture may be given by a patient suffering from any one of several other types of disability of the knee joint. Because of this difficulty mistakes in diagnosis are frequent and a large proportion of the unsatisfactory results of knee-joint surgery are due to this failure.

2. ANATOMY OF THE KNEE JOINT

The knee is the largest, the most important and, in many ways, the most unstable joint in the body. Its strength depends to a large extent upon the ligaments which connect the femur with the bones of the leg, both within and around the cavity of the joint. Surrounding and supporting these ligaments are the muscles of the thigh and leg which, under normal conditions, cushion and protect the ligaments from abnormal stresses. Antero-posterior movement of the femur on the tibia is prevented by the strong cruciate ligaments, extending through the centre of the joint from the femur to the tibia. Complete or partial

The cruciate ligaments

rupture of these ligaments follows only on more severe types of injury, although elongation with subsequent laxity of the joint may be caused by long-continued synovial effusion.

The internal lateral ligament

The internal lateral ligament is fan-shaped, being attached to the internal condyle of the femur at its narrow end and spreading out inferiorly to be attached to the subcutaneous surface of the tibia about half an inch below its articular margin. From the deep aspect of this ligament some fibres are attached to the convex subcutaneous border of the internal semilunar cartilage. In contrast to this arrangement, the rounded cord-like external lateral ligament is attached only to the femur and fibula, and is separated from the external semilunar cartilage by the tendon of the popliteus muscle.

The semilunar cartilages

The semilunar cartilages—wedge-shaped, fibro-cartilaginous structures lying between the margins of the articular surfaces of the femur and tibia—are firmly attached to the upper surface of the tibia by their two extremities, or horns, and in addition are bound to the articular margin of the tibia by short fibrous bands, the coronary ligaments. In spite of their close attachment to the tibia, a certain amount of gliding movement occurs between the cartilages and the head of the tibia, enabling the menisci to follow to some extent the movement of the femur. Normally this latter movement is slight but with general ligamentous laxity there is a considerable increase in this range.

3. DIAGNOSIS AND DIFFERENTIAL DIAGNOSIS

The diagnosis of a cartilage lesion may be a comparatively simple procedure, but, in many instances, a true appreciation of the condition of the joint can be reached only after prolonged and repeated examinations.

(1) Injury of the internal semilunar cartilage

Rotation of flexed knee

The semilunar cartilages are injured only *when a rotating force is applied to* the thigh or leg while the knee is in a position of flexion. In the production of a typical cartilage injury the femur is forcibly rotated inwards, while the foot and leg are fixed on the ground. The story is not always so clear and on occasions there may be no definite history of injury to the joint. As a rule, the

not be straightened through the last 25–30 degrees. On the first occurrence of this so-called locking of the knee it is usually necessary for the patient to obtain medical assistance to regain full movement, but on subsequent occasions he is able, *by twisting the joint in various directions, to correct the displacement.* The restoration of full movement occurs suddenly as the injured cartilage slips back into its normal position. Following the first injury there is, usually, considerable synovial effusion which lasts from 3 to 5 days, but this complication becomes less with each succeeding injury.

Locking

Examination of patient

When the patient is examined soon after the original injury, tenderness is found over the inner aspect of the joint, particularly at two points: the first to the inner side of the ligamentum patellae, and the second at the level of the joint over the middle of the internal lateral ligament. The latter is the more important in diagnosis, for it is caused by strain or rupture of the deep fibres of the internal lateral ligament, which are there attached to the semilunar

cartilage. Accessory aids are used in diagnosis, and of these manipulation of the joint in flexion has proved to be of the very greatest help (McMurray, 1928). Radiographic examination of the knee after the injection of air has been claimed as a valuable aid in diagnosis, and is stated to give useful information in regard to injuries of the cartilages (Cullen and Chance, 1943) (*see* Joints—Arthrography, p. 183.)

(2) Injuries and abnormalities of the external semilunar cartilage

The mechanics of injury to the external cartilage are very similar to those producing a tear of the internal meniscus, the difference being that in this case the rotation is in the opposite direction, the femur being, as a rule, rotated outward on the tibia, whilst the knee is flexed.

Symptoms and signs

The symptoms and signs are very similar to those of injury of the internal cartilage, pain and tenderness and the sense of locking being present on the outer side of the joint. True locking occurs less frequently with injury of the external cartilage and, in addition, slipping of the injured cartilage between the femur and tibia is often accompanied by a loud noise, or click, which is heard with this particular lesion, only when the patient voluntarily extends the knee to the fullest possible extent.

(3) Cyst of the semilunar cartilage

Cystic degeneration may occur in connexion with either cartilage, although the change is found usually on the outer side of the joint. The cyst appears on examination as a smooth, tense structure, projecting outwards, either in front of or behind one of the lateral ligaments. The complaint of the patient is almost invariably of a constant aching pain in the joint, not very severe but increasing with use of the limb. The cyst is more apparent when the knee is fully extended, and the tension in the cyst may be so great that a diagnosis of osteoma may be made if a skiagram is not available. Some difficulty may arise in differentiating between a cyst of the cartilage and an enlarged bursa near the insertion of the biceps tendon; the appearances are very similar, but the complaint of persistent aching, made worse by activity, and the increase in size of the tumour, which occurs when the limb is extended, are sufficient to distinguish the two conditions.

Symptoms and signs

(4) Loose bodies

(a) Aids to diagnosis

Loose bodies in the knee joint may be of fibrous, cartilaginous or osteo-cartilaginous composition, and are usually derived from the articular surfaces, the synovial membrane or the semilunar cartilages.

The symptoms caused by nipping of the loose body between the bones are very similar to those following cartilage injury: *acute pain, locking and unlocking*, followed by synovitis; but the site of locking is not always the *same*, and frequently the loose body can be felt through the skin, usually in the suprapatellar pouch.

Radiographic examination also helps to demonstrate those loose bodies which contain bone, but is of little help when the body is composed solely of fibrous tissue or cartilage.

The diagnosis is comparatively simple; it may be possible to define and palpate the body through the enveloping tissues of the joint and, in many instances, evidence of its presence may be obtained from the skiagram.

(b) Mobility of loose bodies

A loose body may be of a size sufficiently small to pass from one compartment of the joint to another; thus, such a body may at one time appear in the suprapatellar pouch, whilst on another examination it may be found in the posterior compartment of the knee. The larger bodies have not this mobility and, as a rule, remain constantly either in the anterior or in the posterior segments of the joint.

(5) Osteochondritis dissecans

Symptoms very similar to those caused by cartilage injury may be produced by the presence of osteochondritis dissecans. This condition occurs commonly on the internal condyle of the femur near the intercondylar notch (Fig 65). The symptoms are those of indefinite nipping in the joint, rarely a true locking, whilst, in contrast to the cartilage injury, aching (or even pain) is frequently caused by weight bearing.



FIG 65 —Osteochondritis of the knee joint (*By courtesy of Messrs. Edward Arnold & Co.*)

Confirmation of the diagnosis is gained by a good radiographic examination, which shows the irregularity of the articular surfaces and frequently the change in structure of the underlying bony segment.

(6) Rupture of the cruciate ligaments

Although the outstanding sign of this injury is the gross instability of the joint, complete rupture of the anterior cruciate ligament may lead to an erroneous diagnosis of cartilage injury. When a portion of the cruciate ligament still attached to the upper surface of the tibia has become curled on the surface of the bone, nipping between the femur and tibia may produce signs almost identical with cartilage injury. The abnormal antero-posterior laxity of the femur on the tibia is sufficient to differentiate the two conditions.

(7) Adhesions

Following a twist of the joint, adhesions may be formed in the region of the internal lateral ligament or in connexion with the coronary ligament. This strain is followed by a localized haemorrhagic effusion, which is partly absorbed and partly converted into fibrous tissue When, during the movements

of the joint, this inelastic tissue is put under tension, pain follows and the normal voluntary movements of the joint are inhibited.

The diagnosis here rests on the absence of true locking followed by definite unlocking of the joint, on the absence of synovial effusion and on persistent localized tenderness.

(8) Fracture of the tibial spine

This injury usually follows a twist of the joint similar to that causing injury of the semilunar cartilage. The signs are clear: a large haemorrhagic effusion into the joint, inability to extend the limb and great pain if efforts are made to produce this full extension passively. The radiographic appearances make the diagnosis clear.

4. TREATMENT

(1) Treatment of lesions of an internal cartilage

If the patient is seen following the first injury to the cartilage, operation for removal of the injured meniscus is not advisable. If the lesion has occurred on the outer rim of the cartilage, revascularization can take place from the pericapsular tissues, the injured ligamentous fibres may heal and no further disability may be encountered. The tear generally occurs in the substance of the cartilage where no blood-vessels are present, and healing of such tissues has never been demonstrated. It is, however, advisable to treat the first injury conservatively, in the hope that it has taken place in the area in which healing can be expected. If the displacement is repeated, and if there is no decided contra-indication to operation (such as extensive osteoarthritis or obvious ligamentous insufficiency of the joint), removal of the injured cartilage should be undertaken.

(a) Technique of meniscectomy

This operation must never be performed unless the highest degree of asepsis can be guaranteed. The occurrence of infection in a joint cannot be explained away as an unavoidable complication, and the tragedy of ankylosis cannot be excused.

Pre-operative measures

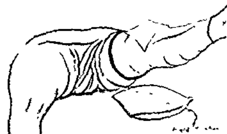


FIG. 66.—Knee prepared for the application of towelling before operation.

The pre-operative preparation of the limb is just as important as the technique used at operation. The whole limb is shaved 2 days before the operation is undertaken; the skin is then prepared with ether soap and methylated spirit, and the limb is wrapped in sterile towels (Fig. 66). On the following day the same routine is followed without a second shaving and, after cleansing again with ether soap and spirit, sterile towels are applied from the groin to the toes and are left in position until they are removed on the operation table.

Preliminary stages of operation

... after elevation ... ared from the use of the tourniquet, which is applied over the dressing ... for the operation time is less than half an hour. The end of the operation table is now let

down, and the patient is placed in a position which allows the leg to hang vertically over the end of the table. A firm sand-bag is placed on this end of the table, directly below the lower end of the thigh and behind the upper end of the tibia. Before the limb is dropped into position over the end of the table, adequate sterile mackintoshes and sheets are used to cover the table, the thigh and the sand-bag, whilst, in addition, the knee area is again painted with methylated spirit, or with iodine, and a covering of sterile batiste, wrung out in methylated spirit, is applied around the joint. This sterile covering of the skin is important; it prevents contamination of the inside of the joint by instruments which would otherwise rest on the uncovered skin.

The incision

The surgeon sits on a stool facing the joint; the stool is of such a height that, in sitting, the surgeon's eyes are just above the level of the head of the tibia An oblique incision of $1\frac{1}{2}$ inches is now made through the covering and skin, from just below the lower and inner border of the patella to a point in front of the tibial attachment of the internal lateral ligament (Fig. 67). With a clean knife the quadriceps expansion is now cut in the same line, and the incision continued through the capsule and the synovial membrane. Two flat Langenbeck's retractors are now inserted into the joint, and the tissues are retracted in order that a view may be obtained of the whole of the anterior compartment of the joint. No forceps of any type are applied to the synovial membrane; this is of the greatest importance, for the injury caused by the application of forceps—even for a very short period—may be followed by a persistent synovitis.

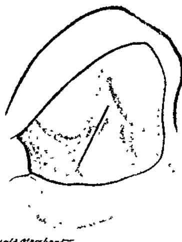


FIG 67.—Incision for removal of internal semilunar cartilage (left knee).

Examination of the joint

It is now possible to see the whole of the anterior compartment of the knee, and any abnormality of the articular surface of the femur, or of the post-patellar pad, can be recognized. In spite of the fact that the anterior portion of the cartilage, which is now visible, may appear to be normal, the surgeon who has made a diagnosis of injury of the internal cartilage should proceed confidently with the operation. *The great majority of cartilage injuries occur in* the posterior half of the meniscus, and the lesion may not be visible from this anterior incision. The object of the operation is the removal of the whole, and not only a portion, of the injured cartilage.

Complete or partial removal

Much discussion has taken place on the advisability of removing only the broken portion of the cartilage, especially when the injury is of the bucket-handle type. If the displaced and partially detached portion only is removed, the final outcome is frequently unsatisfactory. The patient on whom this type of operation has been performed states that the locking which occurred before operation has ceased, but that the knee feels unsteady, especially on rotation, when the femur rides on the rim of cartilage which has been left in position.

Removal of the cartilage: first step

The first step in removal of the cartilage is the definition of its attached edge; to do this a hook is placed around the cartilage (Fig. 68), and an incision is made through the coronary ligament on to the point of the hook. The whole mass of cartilage is then grasped with a heavy forceps and the anterior portion separated from the overlying post-patellar pad and from the head of the tibia, by cutting through the anterior cornu close to the surface of the tibia. While the assistant pulls on the retractors, the marginal rim of cartilage is defined, and is separated by division to the midline, where the deep fibres of the internal lateral ligament are attached to its surface.

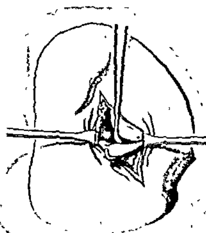


Fig. 68.—Definition of the cartilage by the application of a hook (right knee).

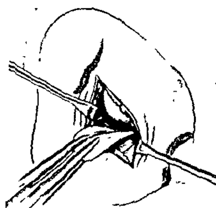


Fig. 69.—Pulling forward the posterior attachment of the cartilage to be removed (right knee)

Second step

The next stage of the operation is extremely important: the deep fibres of the internal lateral ligament must be divided at their cartilaginous attachment. Unless this division is completed satisfactorily at the desired level, complete removal of the cartilage is impossible, for part of the posterior segment must [illegible]

by pulling the anterior portion of the cartilage, by means of the attached forceps, into the centre of the joint, when the division can be completed by a vertical cut. The posterior segment is then easily separated from the capsule by a firm pull of the whole cartilage into the centre of the joint (Fig. 69). As no cutting is required in this section, there is no advantage in the use of any special knife. When the cartilage is fully displaced into the intercondylar fossa its shape is altered by a straight pull, by which its posterior attachment to the head of the tibia is demonstrated. Whilst the cartilage is being pulled [illegible] on its thin [illegible] on and are never produced *in situ* by any type of injury.

Third step

The point of attachment of the posterior horn to the tibia now being visible, the cartilage is there divided by a vertical cut, the *knife being placed on the*

cartilage through the intercondylar space (Fig. 70). By this technique it is possible to remove almost the whole of the cartilage; in fact, it is simpler to divide the posterior attachment of the cartilage in this way than by making a second opening in the posterior portion of the joint.

Closing of the wound

In closing the operation wound the synovial lining should first be sutured, using either plain catgut (which is relatively non-irritating to the joint tissues) or linen thread. In order to diminish the risk of infection of the joint, the catgut must not be handled at any stage, and a careful non-touch technique should be employed throughout the operation. The capsule is then sutured in a similar manner and the skin wound closed with silkworm sutures. In order to prevent any severe effusion into the joint, a large amount of sterile wool is then applied over the dressing and the whole firmly bandaged before removing the tourniquet. The amount of wool is of importance; with a large amount

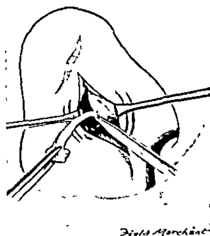


Fig. 70.—Cutting of the posterior horn of the semilunar cartilage through the intercondylar notch (right knee).

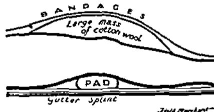


Fig. 71.—Method of dressing and immobilization of the knee after operation.

considerable pressure may be applied to the joint without risk. The limb is placed at rest on a back-splint with a pad under the knee joint (Fig. 71) and the patient is kept in bed for 10 days, when the skin sutures are removed.

After-treatment.—During the next 10 days the patient walks about, still wearing the back-splint, and at the end of this period the splint is replaced by a bandage around the joint, the upper level of the bandage being the upper margin of the patella. Quadriceps exercises should be practised while the splint is being worn, and the surgeon need not fear any great loss of power in the muscles of the thigh, so long as the operation has been carried out gently and without undue trauma to the joint. Restoration of the full range of movement in the joint can be left safely to the patient's own efforts. On removal of the splint the patient can usually bend the knee to an angle of 50–70 degrees, and after a week the remainder of the movement returns. Voluntary exercises of the quadriceps should be encouraged during the period of rest, and later on massage of the same muscle soon restores full power in the limb, a return to full activity being possible 3–4 weeks after the operation.

This routine of after-care has been arrived at after an extended experience of treatment by early active joint movements, and the results obtained by it have been so much better that the method of early activity has been abandoned.

(b) Disability following incomplete removal

After the incomplete removal of a cartilage, discomfort and disability are frequently experienced in the knee. Locking is not then a feature, but the patient usually complains of a sudden repeated nipping in the joint, causing the limb to feel momentarily powerless. Effusion may or may not follow these occurrences but, as a result, athletic activity cannot be carried on successfully. The patient, as a rule, is unable to define the point at which nipping occurs but, on examination of the joint by manipulation, the same sensation

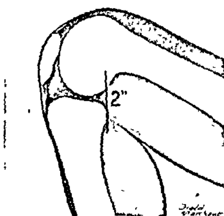


FIG. 72.—Incision for removal of posterior end of semilunar cartilage or for removal of loose body in the posterior segment of the joint (right knee).

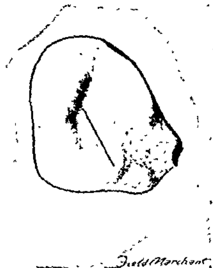


FIG. 73.—Incision for removal of external semilunar cartilage (left knee).

of nipping and also a distinct click can be reproduced and localized in the posterior compartment.

(c) Removal of the remaining posterior segment

Removal is most readily effected through a postero-internal incision, made with the knee bent to a right angle (Fig. 72). With the foot lying on the table against the patient's other leg, a vertical incision, 2 inches in length, is made behind the internal condyle of the femur and the upper and inner border of the shaft of the tibia. The centre of the incision lies just above the posterior margin of the articular surface of the tibia; in this position of flexion the inner hamstrings fall backwards and the incision can be continued safely through the tissues until the bulging capsule of the joint appears in the wound. The capsule is now opened in the same line, when the posterior end of the cartilage can be readily defined from its posterior attachment to the point at which it has been divided in the previous operation. After the margin has been freed from its capsular attachment, the stump of the cartilage is held in strong forceps and drawn into the wound, where its posterior attachment is divided with a knife or tenotome.

(2) Removal of the external cartilage

The steps of this operation are almost identical with those employed in removing the internal cartilage. The incision is of the same length, passing

obliquely down from the lower and external border of the patella towards the external margin of the upper articular surface of the tibia (Fig. 73). The separation of the cartilage from its attachments is no more difficult than on the inner side of the joint, and the division of its posterior attachment is completed in the same manner. As in the case of removal of the internal cartilage, post-operative disability may occur if the whole of the cartilage has not been removed; the symptoms are similar, and a decision as to the condition is made by manipulative examination.

Removal of the posterior segment of the external cartilage

With the knee again flexed to a right angle, a vertical incision of 2 inches is made through the skin in a line between the biceps tendon and the external lateral ligament. When the incision is deepened, the bulging capsule of the knee becomes apparent, and when the joint is opened a smooth white rounded structure is seen, extending from before backwards at the level of the upper surface of the tibia. This is the tendon of the popliteus muscle and, because of its appearance and its situation, mistakes have frequently been made, and removal of the tendon has not improved the condition of the knee. In order to see and deal with the cartilage, the tendon is first hooked upwards, when the cartilage can be seen lying in its normal site over the posterior portion of the tibial articular surface. Its removal is a comparatively simple process after its posterior horn has been divided close to the tibia.

(3) Treatment of cysts of the cartilage

Either aspiration of the fluid content of the cyst or local removal has always led to recurrence and to a continuation of the symptoms. The only satisfactory treatment is removal of the whole of the cartilage from which the cyst is derived.

Technique

The operation of removal should be carried out with exactly the same technique as that employed for removal of an injured meniscus. Some surgeons believe that removal of the cartilage with the whole cyst and cyst wall is a necessity; in my experience the essential point of the operation is removal of the whole of the affected cartilage even though a portion of the cyst wall may be left *in situ*. This leaves a better knee joint and does not involve the long period of incapacity and convalescence which follows the more extensive operation. When the operation is completed in the manner suggested the synovial membrane is not so widely removed. Closure of the wound is

of treatment used following the operation. It is claimed that early active movements of the joint prevent stiffness, maintain the group of muscles at almost their normal strength and allow of earlier activity. In my experience, after the unavoidable trauma of the operation, the joint requires rest above everything else, in order to prevent synovial effusion and even haemarthrosis. If the operative technique is gentle, if forceps have not been applied to the synovial lining, and if the line of sutures in the synovia is satisfactory, stiffness of the joint is not to be expected. In spite of the presence of serious changes in the joint, removal of a body which is causing repeated lockings is justifiable, the subsequent function of the limb being thereby improved

(4) Removal of a loose body

(a) Technique

The approach must depend upon the situation of the loose body. If the skiagram indicates that the body is of relatively large size and is situated in the posterior compartment of the knee, it is evident that the incision for the opening of the joint must be made in the same line, and with the same technique as that used for removal of the posterior end of the internal or external cartilage, the site of the incision depending upon the clinical signs, history and radiographic appearance. When the body is small, and has been localized in the anterior portion of the joint or in the suprapatellar pouch, the incision for its removal should always be made on the outer side of the patella. A vertical incision is used, extending from the level of the middle of the patella upwards for 2 inches (Fig. 74); the knee is then held flexed to an angle of 10 degrees, and the joint is covered posteriorly and laterally with a sterile towel. With his hands on the outside of this towel the surgeon then massages the knee carefully from every angle, in an effort to force the loose body into the operation field. (Fig. 74). The simplicity of this procedure is astonishing; not only is it possible to bring the loose body from any portion of the front of the joint but, if small in size, the body can be brought from any part of the posterior compartment.

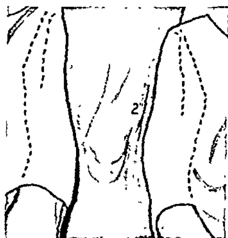


FIG 74 —Showing incision, and finding of loose body in the right knee joint.

(b) After-care

The after-care of the patient is identical with that used after removal of the cartilage, if the articular surfaces are healthy; if, however, severe arthritis is present in the joint, the stabilizing splint should be omitted and a limited range of movement allowed from the first.

(c) Prognosis

With a single loose body in a knee joint, the articular surfaces of which show little sign of arthritic changes, the prognosis for the function of the joint is favourable, but when several bodies are present, and especially when the skiagram indicates severe joint destruction, the outlook for the future is poor.

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JOINTS—TUBERCULOSIS

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1. DEFINITION

207.] Tuberculosis of a joint implies invasion by the tubercle bacillus, and also the destruction of the adjacent bone.

2. AETIOLOGY

Two types of tubercle bacilli produce disease in man, the human and the bovine. "Open" pulmonary lesions are the source of the human type, infection being conveyed by droplets or dust. Milk is the source of the bovine type.

In an investigation carried out at the Orthopaedic Hospital, Oswestry, Shropshire, upon cases of bone and joint tuberculosis, 100 samples of pus from which the tubercle bacillus was obtained were typed, and in all the human bacillus was isolated. (The typing was done by Dr. Rhodes in the Pathological Department of the Shrewsbury Royal Infirmary.) Even in childhood, when tuberculous pus does not often appear, the human type is obtained. This would suggest then, that correction of the milk supply alone will not eradicate the disease.

Route of infection

Once in the tissues, the bacilli reach the lymphatic glands, and cause there the usual typical pathological changes. The infection may proceed no farther, and may lie dormant. When the resistance of the body is lowered a "flare-up" occurs, perhaps many years later. The glands soften, a neighbouring blood-vessel is eroded and so the blood stream is infected. Another route to the general circulation is via the pulmonary capillaries, following inhalation of a massive dose of the bacilli, and sometimes spread is by the lymphatic duct.

The joint infection may occur as a late manifestation of a general tuberculous infection, and sometimes follows on operation for tuberculous disease elsewhere, for example, nephrectomy.

Age incidence

Tuberculosis of bones and joints occurs at any age, from infancy to old age. It is a mistake to look upon it as a disease of childhood.

3. PATHOLOGY

A joint is nearly always invaded from the blood stream, but the spine may become affected by direct lymphatic spread from the rich lymphatic area in the dorso-lumbar region, and the hip joint directly from a psoas abscess.

Blood-stream infection of a joint may occur in one of two ways: by direct implantation of the bacilli into the synovial membrane, commonly seen in the knee joint in children, or as an initial infection of the adjacent bone, spreading thence to the joint (Fig. 75). The articular cartilage is destroyed and replaced by a tuberculous "pannus" of granulation tissue. The articular ends of the bones are destroyed; ligaments are softened and stretched and, if the joint is not protected, dislocation occurs. The classical example is the "wandering acetabulum" of the hip joint (Fig. 76).

Destructive changes in the joints



Fig. 75.—Tuberculous focus in neck of femur. Danger of spread to the joint

Tuberculous pus

The liquefied debris forms tuberculous pus, and appears in the tissues as a cold abscess. This pus soon breaks out of the joint, forming an extra-capsular abscess, and will quickly break through the skin if not held in check by aspiration. Tubercle bacilli can be demonstrated in the pus, and although bacilli are present, repeated examinations of the pus by culture or animal inoculation may be necessary to give a positive result.



Fig 76—Pathological dislocation of the hip—the "wandering acetabulum".

The healing process

Healing takes place over a period of many years, but the joint is destroyed and filled by granulation tissue which slowly organizes into an avascular fibrous tissue containing islets of tubercle bacilli which lie dormant for many years. Reactivation occurs because of poor general health, intercurrent illness, or sometimes after an injury.

Sound and unsound fibrous ankylosis

The fibrous ankylosis that occurs is called "sound" if the joint is clinically completely stiff, with no detectable movement. The usual end-result, however, is an "unsound" fibrous ankylosis To make such a joint "safe" an arthrodesis must be carried out. Bony ankylosis may occur spontaneously, especially in the sacro-iliac joint. In other joints ankylosis follows only a secondarily infected sinus, staphylococci being a well-known stimulant of osteogenesis Operative treatment is designed to secure a bony ankylosis when this has not occurred spontaneously.

Bony ankylosis

4. CLINICAL PICTURE

(1) Diagnosis

Relationship to trauma

The history of an injury is important and may be misleading. Patients often ascribe their illness to an injury, and because of this a tuberculous synovitis may be mistaken for a traumatic effusion. For the purposes of litigation it is usually said that although an injury, *per se*, could not cause tuberculosis *of a joint, yet* it can lower local resistance sufficiently to allow tubercle bacilli to settle and thrive in *the damaged tissues.*



FIG 77.—Tuberculosis of the shoulder Note *erosion of head of humerus and cavity in* greater tuberosity. The glenoid fossa has been *involved more than is usual in a caries sicca.*

(2) Signs and symptoms

(i) *Shoulder.*—Moderate subdeltoid swelling is present together with pain on movement. There is wasting of the muscles of the shoulder girdle and as the head of the humerus crumbles (*"caries sicca"*) (*Fig. 77*), pathological subluxation occurs, so [illegible] shoulder, [illegible] th [illegible] or [illegible] us tracking along the synovial sheath surrounding the long head of the biceps may form a *swelling at a considerable distance from the joint.*

(ii) *Elbow.*—There is white swelling, pain and spasm with limitation of movement. Abscesses and sinus formation are common because the joint is superficial (Fig. 78).



FIG 78.—Tuberculosis of elbow. Note extensive bony destruction and soft-tissue thickening.

(iii) *Wrist.*—Characteristic *spindle-shaped swelling of the* joint is seen, together with palmar flexion deformity. Abscess and sinus formation occur early because the joint is largely superficial. The outlook is exceedingly grave because multiple secondarily infected sinuses may be a source of danger to life, or may result in such severe crippling of the hand that amputation becomes necessary.

(iv) *Hand.*—The metatarsals and phalanges are affected. Tuberculous dactylitis is common in children, with characteristic spindle-shaped swellings of the short bones, frequently healing spontaneously without abscess formation.

This causes shortening of the fingers. The metacarpo-phalangeal or inter-phalangeal joint may be involved, and in adults this may be the initial focus in the digit (Fig. 79).

(v) *Hip.*—The early symptom is stiffness rather than pain. Movements are limited in all directions, and there is wasting of the thigh and buttock muscles. Characteristic abduction-external rotation deformity occurs in the early stages, causing an apparent lengthening of the limb. In the later stages there is an adduction-internal rotation deformity, with true shortening when pathological subluxation is threatening. Abscesses occur late, because this is a deep joint, but they do occur very frequently even in children. They usually appear over the trochanter but are seen also in the femoral triangle or in the adductor region, and occasionally they may track down into the thigh.

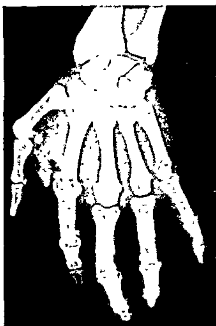


FIG. 79.—Tuberculosis of the metacarpo-phalangeal joint of index finger in an adult.

(vi) *Knee.*—In childhood there is nearly always "white swelling" because the infection begins in the synovial membrane. Caseation and pus formation do not occur. In adults there is usually marked swelling, with pain, muscle spasm, raised local temperature and the early formation of an abscess. There is also gross wasting of the limb. Extra-capsular abscesses form and may burrow far into the thigh. The condition may be very chronic and difficult to diagnose. Difficulty is sometimes experienced in differentiating the sub-acute form from pyogenic arthritis. The knee joint is very frequently involved and lends itself to the special aids to diagnosis; the bed-rest test, inguinal gland biopsy, the erythrocyte sedimentation rate estimation and, in certain circumstances, biopsy of the synovial membrane may be necessary (*see* p. 227).



FIG. 80.—Tuberculosis of the ankle joint with destruction of the astragalus and intense decalcification of the adjacent tibia.

(vii) *Ankle.*—As this is also a superficial joint, white swelling can be seen, and there is pain, muscle spasm and considerable wasting of the calf. Abscesses and sinuses are common (Fig. 80)



FIG. 81.—Tuberculosis of the metatarsal bone in a child, with abscess and thin overlying skin which will soon form a sinus.

(viii) *Foot.*—In childhood the disease tends to pick out one tarsal bone. Abscesses occur (Fig. 81). In the adult the disease is often more widespread throughout the tarsal joints (Fig. 82). Metatarsals, phalanges and their related joints are also affected, giving the usual signs. Infection of metatarso-phalangeal joints frequently forms sinuses in the webs of the toes.

(ix) *Effects on the general health.*—Because tuberculosis is a general systemic disease and the joint lesion only a local manifestation of it, there is malaise, with loss of weight and of appetite. Night sweats occur, and the erythrocyte sedimentation rate is raised.

(3) Complications

(a) *Cold abscess*

The formation of a cold abscess

Abscess formation is a comparatively late manifestation of the disease, and in a deeply placed joint such as the hip, the appearance of an abscess under the skin means that the disease has been present many months. The pus, which in the earlier stages contains polymorphonuclear leucocytes may form in large quantities. The abscess wall ultimately becomes thick, fibrous and calcified. Sooner or later abscesses appear in any joint which is involved—except in caries sicca of the shoulder. In superficial joints abscesses rapidly become extracapsular, and are therefore quickly detected.

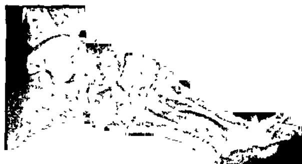


FIG 82.—Tuberculosis of the astragalo-scaphoid joint in an adult. Note the extensive destruction.

(b) *Sinus formation*

Secondary infection

A sinus must always be regarded as a serious complication, because of the mortality from secondary infection. When secondary infection develops, osteomyelitis or acute suppurative arthritis is likely to occur, and must be treated by adequate

drainage and chemotherapy. If the secondary infection is mild, the usual end-result is a bony ankylosis (Fig. 83). Unfortunately, as in pyogenic osteomyelitis, recurrences in later life may be frequent.

(c) Multiple lesions

Frequently more than one joint is involved. Any and every combination occurs, and in many there is a history of tuberculous ribs or dactylitis years before. The nursing of these patients is a major problem.

(d) Deformities

Pathological dislocations, subluxations and contractures are not uncommon. These have been mentioned under the various sections and their treatment is described later.



FIG. 83.—Old healed tuberculosis of the foot with sinuses. Note the solid bony ankylosis

(e) Involvement of other systems

These may become involved in addition to the primary and skeletal lesions. They include the lungs, genito-urinary tract, tendon sheaths and the skin In children tuberculous otitis media and mastoiditis are common, and tuberculous meningitis is responsible for a percentage of deaths.

5. SPECIAL AIDS TO DIAGNOSIS

(1) Radiological appearances

The earliest change, which takes about 6 months to develop, is a diffuse decalcification of the bones close to the affected joint—a result of hyperaemia causing absorption of the lime salts Widening of the joint space may occur, and is due to the effusion, but this is not a reliable early sign. Alteration of the shadow thrown by the soft tissues is of great value, for example, an effusion into the hip joint or knee joint causes a ballooning of the capsule. Thickening of the synovial membrane and of the periarticular tissues from oedema is shown by their increased density. In acute pyogenic infections of joints there are no radiographic changes in the first few weeks, but later there is a rapid decalcification and woolliness of the joint outline. In a specific arthritis from a gonococcal infection the diffuse decalcification takes about 6 weeks to develop. A disuse decalcification causes a stippled appearance of the bones.

Earliest change

Differential diagnosis

When the infection begins near a joint, a cavity forms, and may contain calcified debris (Fig 84). The earliest radiographic sign of this is an erosion of the bone trabeculae.

In later stages the outline of the joint becomes hazy or "feathery", and finally all stages of destruction are shown, until the joint becomes completely disintegrated and loses its identity as an anatomical unit (Fig 85).

Later stages

A skiagram may show the outline and size of a calcified abscess cavity, and this may be of importance in deciding what form of arthrodesis to carry out—

for example, in the hip an abscess sometimes forms under the femoral neck, and contra-indicates an ischio-femoral arthrodesis.

Healing stages

The healing stages are shown by the improved lime content of the bones and the improvement in the joint outline, but osteogenesis does not occur in a pure tuberculous infection as it does in a pyogenic one.



FIG 84 —Two tuberculous foci in the foot. Note the extensive cavity in the first metatarsal, and the small cavity in the head of the os calcis

(2) Special investigations

(a) *Erythrocyte sedimentation rate*

In monarticular tuberculous arthritis the average fall in the first hour is usually about 20–30 millimetres (Westergren). This steadily becomes less with suitable splinting. When large quantities of pus are being formed, however, the reading in the first hour may be as much as 100 millimetres.

A steady diminution in the erythrocyte sedimentation rate means a favourable response. In the subacute stage of the disease normal figures may be obtained. The chief value of the erythrocyte sedimentation test is as an indication of the progress of the disease.

(b) *Tuberculin tests*

A positive result is inconclusive, but a proved negative test rules out the diagnosis of tuberculosis.

(c) *Gland biopsy*

This is helpful in the hip joint and knee joint. The lymphatic drainage of the hip passes through a nest of glands around the external iliac vein, and that of the knee through the inguinal glands. It is unnecessary to carry out this examination in every case, but it should be done



FIG. 85 —Late stage of tuberculous knee. Note extensive bony destruction and the hazy outline and texture of the bones.

when all other tests have proved negative, and when radiographic changes remain indefinite.

(d) Diagnostic aspiration

Every abscess should be aspirated at least once for diagnostic purposes, and this may confirm the diagnosis. Animal inoculation or culture of the fluid is usually necessary.

(e) Arthrotomy

It is unwise and usually unnecessary to perform a synovial biopsy in the presence of acute disease, because a sinus may develop before an arthrodesis is required. It is tempting to explore a superficial joint such as the knee, but the temptation should be resisted, and the exploration should never be done in children. The superficial joints lend themselves to this form of examination.

(f) Bed-rest test

In many patients this is the most satisfactory and reliable of all tests and one which has to be used frequently for diagnostic purposes. The affected joint is placed at rest, and if the effusion rapidly subsides and does not reappear on movement, tuberculosis can be ruled out, and the condition is probably a mild toxic or traumatic arthritis. A gonococcal arthritis also responds well to rest and graduated exercise but requires a period of some weeks. The swelling of a tuberculous joint, though diminishing somewhat with rest, never completely subsides, and with movement the swelling, pain and muscle spasm quickly return.

(g) Serological tests

The Wassermann and Kahn reactions should be done as a routine. The gonococcal complement fixation test is not so reliable, because of the wide range of conditions which also give a positive result.

6. DIFFERENTIAL DIAGNOSIS

A tuberculous joint may have an acute onset, resembling a pyogenic infection; but the onset may be insidious, and difficult to distinguish from many obscure affections of joints.

(1) Acute pyogenic infection

Acute suppurative arthritis

Acute suppurative arthritis of haematogenous origin (staphylococcal, streptococcal and pneumococcal) is an extremely acute illness. The onset is sudden, with high temperature and toxaemia. The affected joint is exquisitely tender, red and hot, and there is much pain and spasm. The diagnosis is made by culture of the bacteria from the blood or from the joint fluid.

(2) Specific arthritis

Gonococcal arthritis

Gonococcal arthritis deserves special mention because of its close resemblance to tuberculosis in its early stages; the history of a urethritis may not be forthcoming. The bed-rest test is of great value here. A gonococcal joint responds to rest and penicillin. The gonococcal flocculation test, if strongly positive, is helpful.

Syphilitic arthritis

Syphilitic arthritis occurs in both the congenital and acquired forms. The joint enlargements are usually multiple; they occur in the new-born as well as in later life and are associated with other soft tissue and skeletal changes.

In older children one joint alone (such as the knee) may be involved and the condition closely resembles tuberculosis. In such a case the Wassermann reaction is positive, and it may be permissible to carry out a synovial biopsy to show the absence of tuberculosis.

Acute rheumatic fever in children causes a mild fleeting arthritis and pyrexia. The multiplicity of joints involved and the fleeting character of the symptoms *distinguishes the condition from tuberculosis.*

(3) Neuropathic arthritis

Disease of the nervous system, commonly tabes dorsalis or syringomyelia, causes a chronic enlargement of joints originally described by Charcot and known by his name. The joint affected is usually painless and remarkably unstable. The skiagram shows absorption of the joint surfaces, sometimes of extreme degree, but there may be in addition much irregular formation of new bone.

(4) Non-specific arthritis

A *toxic arthritis due to sensitivity to various substances such as bacterial* toxins, drugs and sera may cause monarticular or multiarticular effusions usually of a temporary nature and not associated with any thickening of the periarticular tissues. There may or may not be a generalized systemic upset, and skiagrams are negative. When one joint alone is involved, tuberculosis may be ruled out by discovering the cause of the condition and by negative results to specific tests, but there are many joint effusions of which the diagnosis remains obscure.

Urethritis with arthritis

A fleeting synovitis occurs which is associated with a non-specific urethritis. This rapidly responds to rest, does not recur on movement and all the tests are negative. The diagnosis is made by proof of the non-specific nature of the urethritis. The arthritis and urethritis are sometimes associated with a conjunctivitis, when the triad is known as Reiter's disease.

(5) Chronic rheumatic joints

Rheumatoid arthritis

Rheumatoid arthritis may have an unusual onset; it may begin as a monarticular arthritis in one large joint such as the knee, before any of the usual changes have occurred in the small joints of the hand and fingers. The patient is usually a female, in her thirties, the clinical appearance and progress of the joint condition closely simulates tuberculosis, and there is an increased erythrocyte sedimentation rate. A synovial biopsy would show the presence of a non-specific inflammation, and the appearance later of enlargement of the *small joints of the hands and fingers establishes the diagnosis*; in Still's disease an effusion may appear in one knee long before any other joint is involved.

(6) Osteoarthritis

This condition may cause a *recurrent synovitis and symptoms over a period* o [illegible] ir [illegible] degenerative changes as seen on a skiagram, may cause [illegible] joint has been observed under traction for a few months, when further skiagrams will show more destruction if the condition is tuberculous.

(7) Osteochondritis

This gives the signs and symptoms of a mild synovitis or arthritis. Before the use of x-rays, osteochondritis was frequently mistaken for tuberculosis, the classical example being Perthes's disease of the hip joint.

(8) Perthes's disease of the hip

The symptoms are those of a mild recurrent arthritis occurring in childhood. The general health is not affected; pain and spasm are never severe and pass off quickly after a short period of rest. The child usually remains on his feet, and movements of the joint are hardly restricted. The diagnosis is established by skiagram, which shows characteristic changes depending on the stage of the disease. The femoral head is sclerosed early, and fragmented later as revascularization occurs (Fig. 86). At the onset of the disease, however, there are no radiographic changes, and apart from spasm of the hip there is little to be made out.

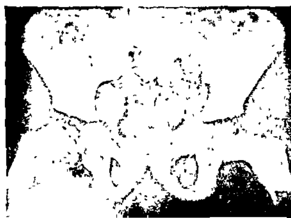


FIG 86.—Skiagram of Perthes's disease of the hip Note the fragmented appearance of the upper femoral epiphysis

(9) Osteochondritis of the tarsal navicular

Köhler's disease

Köhler's disease also occurs in childhood. Clinically there is swelling, tenderness and possibly raised local temperature. The skiagram shows the sclerosed or fragmented appearance of the bone, and this may closely resemble tuberculosis. If the condition is tuberculous sooner or later an abscess appears, from which the bacillus can be cultured (Fig. 87); if osteochondritic, steady improvement occurs with plaster fixation.

Freiberg's disease *Kienböck's disease* *Panner's disease*

Freiberg's disease of the second metatarsal head and Kienbock's disease of the carpal lunate in adults are not likely to cause difficulty with diagnosis because of the characteristic radiographic appearances, but Panner's disease of the second and third metatarsal heads, in which chronic swelling of the affected joints occurs and the skiagrams show absorption of the bones without new bone formation, may cause difficulty. In this condition there is remarkably little pain and spasm and pus does not form. A biopsy may be required to establish the diagnosis.

(10) Haemophilic joints

Haemophilia

In this affection, recurrent haemorrhages occur into the joints, especially in the hip and knee, and not necessarily after an injury. Though the appearances of the joint may resemble tuberculosis, the family history and the patient's past history will usually suggest the correct diagnosis.

(11) Protrusio acetabuli

Otto's pelvis

There are many causes of this condition, but that associated with the name of Otto causes a unilateral or bilateral "crumbling" of the hip joints

Skiagrams show a uniform loss of joint space, and a deepening of the socket. When this occurs in one hip the clinical picture may resemble tuberculosis. The radiographic changes are not the same, however, and abscesses do not form. This condition responds to rest and traction and although the acetabulum is deepened a useful range of movement can sometimes be preserved. The cause is probably chronic infection, but there may be the history of an injury. The condition may also follow an acute arthritis.







Fig 87.—Example of tuberculosis of the tarsal navicular in a child. There was close resemblance to osteochondritis of this bone but an abscess containing tubercle bacilli ultimately appeared.

(12) Endocrine disorders

In young women effusions into the knee joint, which appear to have some relationship to menstruation, sometimes occur. The symptoms are never acute and there is no restriction of movement. Treatment is unsatisfactory but the condition does not appear to progress or to cause any undue disturbance of the general health.

Slipped upper femoral epiphysis

Slipped upper femoral epiphysis is probably caused by combination of trauma and endocrine disorder. There is an unmistakable clinical and radiological appearance.

(13) Tumours

(a) Synoviomas

Synoviomas or endotheliomas arise from the synovial membrane. The history runs a chronic course and in the early stages may resemble an internal derangement, especially if there has been an injury. The knee is a common site. Clinically there is a doughy swelling as a result of the thickening of the synovial membrane, closely resembling tuberculosis. Skiagrams show a diffuse decalcification of the bone ends. The diagnosis is made usually only after a synovial biopsy.

(b) Sarcomas of bone

Sometimes these tumours arise near a joint, especially at the lower end of the femur and upper end of the tibia. In the early stage they may resemble tuberculosis, and cause recurrent effusions into the related joint, with pain

and muscle spasm which are unrelieved by rest in bed. Skiagrams at an early stage may show little change except a patchy decalcification. As time passes, expansion of the bone can be felt in the juxta-epiphyseal region, and periosteal new bone and destruction of the cortex are visible in a skiagram.

(c) *Bone cysts*

These occur either singly or in fibrocystic disease of bone. The femoral neck is a common site; a biopsy is usually necessary (Fig. 88).

(d) *Synovial chondromatosis*

This occurs particularly in the knee. Clinically there is a persistent swelling of the joint, without much restriction of movement, unless a loose body causes locking. The skiagram shows *multiple small deposits of calcium and a general decalcification.* A *biopsy reveals* large numbers of cartilaginous loose bodies in the joint.



Fig. 88.—Tuberculous tibia proven by exploration. Child too young for an osteoclastoma. This might have been a multilocular cyst.

(14) Trauma

Internal derangements and loose bodies can usually be distinguished by the history and the clinical and radiological findings. Osteochondritis dissecans of a femoral condyle causes recurrent effusions and pain before the loose body has separated, but *the nature of* the condition can usually be *seen on the skiagram.* Hyperaemia as a result of injury, if followed by rest, causes a patchy "disuse" decalcification which slowly disappears as the function of the joint returns to normal.

7. PROGNOSIS

It is uncommon to get painless, safe and useful movement in a joint following tuberculosis. It does occur in the hip and knee, especially in childhood. A full range of movement is never obtained and such a joint is rarely safe, as will be seen later. Although *accurate figures cannot be given, it is doubtful whether* more than 3 per cent get a movable joint. It rarely occurs in an adult. Prognosis *as regards life should always be guarded until the disease becomes* stabilized. The presence of a sinus greatly increases the risk, and the appearance of *the disease in other systems* is all too common.

Range of movement

8. TREATMENT

(1) General

As Phillips said, tuberculosis is a "fever diluted by time". The general treatment overshadows all else, and operative treatment *is only an incident.*

The joint a local manifestation of tuberculosis

The patient with a tuberculous joint must be admitted to an open-air country hospital, otherwise failure in treatment may occur. It is common to see a wasted patient with a swollen joint admitted to such surroundings, and an almost miraculous change for the better occur in this environment. With open air, sunlight (which must be graduated at first) and a generous diet the patient rapidly gets a feeling of well-being, his general metabolism is stimulated, his peripheral circulation improves and he can fight his disease. Hand in hand with the improvement in the general condition, the joint swelling subsides, abscesses are absorbed, and the local disease soon becomes quiescent.

An open-air ward



FIG. 89.—An open-air ward. Note the wheel-beds.

An open-air ward means a ward with only three walls. (Fig. 89). It is not possible to heat such a ward, and in winter the patients are kept warm by suitable bed-making and hot-water bottles.

As most patients are long-stay cases, the beds should be on wheels, so that the patients can be taken round the grounds, and to the hospital canteen by their relatives and friends; thus boredom is avoided.

Adolescent problems

Occupational therapy is important especially in the adolescent after leaving school. Every effort must be made to keep the patients employed.

In hospital, children continue their education, which is provided and supervised by the Ministry of Education.

(2) Splinting

(i) *Hip.*—The Jones abduction frame consists of an iron framework and padded leather saddle, with a groin strap placed on the side opposite the lesion (Jones, 1904). The iron frame can be adjusted for abduction, and the saddle extends from the nape of the neck, bifurcating at the hips for each thigh, and reaching to the head of the tibia. Here the tibia must be supported slightly flexed, to avoid subluxation of the knee joint (Fig. 90)

The lower limbs are "steadied" by means of skin extensions with tapes tied to the ends of the splints.

[illegible] support by [illegible]

(Thomas, 1890) (Fig. 91).

(iii) *Ankle and tarsal joints.*—A closed padded plaster, with windows for sinuses if necessary, forms a suitable splint. The disadvantage of a closed plaster is that the air is prevented from stimulating the skin and so improving the peripheral circulation. The crab splint, a malleable metal frame made

from a cast of the foot, overcomes this. It is a difficult splint to fit, however, and requires much attention.

(iv) *Shoulder.*—A plaster spica is applied with the shoulder abducted to 80°, externally rotated 25°, and with the elbow in the forward plane of the chest. The spica must grip the pelvis. A metal-framework splint (Littler-Jones) can be used, but is difficult to fit and to maintain. It is useful, however, in the presence of sinuses. In the later stages when the shoulder is awaiting arthrodesis, a collar-and-cuff sling is sufficient.

(v) *Elbow.*—A plaster case is applied in the optimal position —just below a right angle in most cases. Later a block leather or other lacing splint is applied.



FIG 90.—Patient with tuberculosis of the hip on a Jones abduction frame.



FIG 91 —Tuberculous knee with sinus in a Thomas knee bed splint. Plaster splints to give additional support because the condition of knee was so acute

(vi) *Wrist.*—A closed padded plaster or metal-framework cock-up splint is used, with the fingers free.

(vii) *Metacarpals and phalanges.*—For metacarpals a small cock-up splint of metal or plaster can be applied. The fingers should be free to avoid stiffness. In children tuberculous phalanges require small individual finger splints; in the adult amputation is the best treatment.

(viii) *The sterno-clavicular joint.*—This joint will be mentioned here because of the unusual features attached to it. It is not often affected alone and is usually seen in a patient with other skeletal lesions. The joint becomes chronically enlarged and a sinus often forms. Treatment consists of applying a collar-and-cuff sling until the disease is quiescent and later excising the joint. A bony ankylosis does not occur, probably because of the difficulty of fixation. The sound fibrous ankylosis which

follows the excision appears to do no harm, and is not aggravated by movements of the shoulder.

Benefit of the framework type of splint

The benefit of the iron-framework type of splint is that the air and sun can get to the part and stimulate the peripheral circulation. A closed plaster-of-Paris case prevents this. At the same time over-splintage is to be avoided. It is true to say that, especially in infants, no splint gives absolute immobilization, and indeed it is doubtful whether this is desirable. The affected joint is "steadied" and all other joints should be kept supple. Quadriceps drill and foot exercises should be practised by patients who are recumbent on frames. The iron-framework splints have proved to be a happy medium between rest and movement.

Indications for progression of treatment

The next problem to decide is how long a patient is to remain in his splint. The general health of the patient must be excellent, and the joint quiescent. A series of skiagrams must show not only that no further destruction has occurred, but also that there is a steady improvement in the lime content of the bones This may take a number of years. An ambulant splint is then applied if arthrodesis is not to be carried out immediately.



Fig. 92.—Tuberculosis of the knee in later stages with walking caliper fitted. Note support to prevent knee hyperextending The ends of the Thomas splint are sawn off and turned into slots in the heel of the boot.

(i) *Hip.*—A patten is placed on the foot of the unaffected side and the patient is allowed to walk in a plaster spica, using crutches, but pattens are not altogether satisfactory because the patient "stumps" down on the foot of the affected side. Direct weight-bearing therefore is allowed as early as possible Later a lacing block leather spica is applied, but has the disadvantage that, as all tuberculous hips tend to adduct, the edge of the block leather cuts into the adductor muscles and causes considerable wasting. A sound arthrodesis is much better. If an arthrodesis is carried out successfully the minimal time of treatment of a tuberculous hip is 5 years, though continual stay in hospital is not necessary.

(ii) *Knee.*—A weight-relieving caliper is applied (Fig 92). In the adult this would be followed by an arthrodesis which, if successful, allows freedom from all splintage.

(iii) *Ankle and foot.*—A below-knee walking plaster is applied, and later a below-knee double iron with drop-foot stops.

(iv) *Shoulder.*—If a sound fibrous ankylosis occurs, the splints are removed and scapular movement is encouraged. Later, if there are no sinuses, an arthrodesis is carried out.

(v) *Elbow.*—A block leather or other form of lacing splint is applied and later an arthrodesis is performed.

(vi) *Wrist.*—Treatment is the same as that for the elbow.

(vii) *Metacarpals and phalanges.*—These frequently require no splintage

unless the small joints are involved. A small metal splint is all that may be necessary.

(3) Treatment of cold abscess

If the abscess is small, and not enlarging, and the joint is suitably splinted, then apart from a diagnostic aspiration the abscess should be left alone.

If the abscess is large or enlarging, repeated aspiration through healthy tissues should be carried out until the abscess remains flat.

The application of external pressure will not prevent an abscess from filling and enlarging. Rapid filling of an abscess after aspiration indicates acute disease, and the pus will contain large numbers of polymorphonuclear leucocytes. This process is not influenced by external pressure.

Every effort must be made to avoid sinus formation. The injection of drugs into the abscess cavity has not met with much success, nor in the case of a large abscess has clearing-out and curettage, followed by closure of the skin, unless the abscess cavity is of an extremely chronic nature. When the pus is very thick, and blocks the aspirating needle, it is permissible to make a small incision with a scalpel, express the thick material and close the incision with one silkworm gut suture.

(4) Treatment of sinuses

Prevention of secondary infection

Once a sinus has formed much can be done to prevent serious secondary infection by careful "non-touch" technique in dressings, and, when cleansing the skin, by taking care to swab in a direction radiating from the sinus. Dressings should be infrequent and masks must be worn.

(5) Treatment of other complications

Correction of deformities of the hip

(i) *Hip*.—Traction in the line of the deformity usually overcomes rigid adduction deformity. When there is a pronounced flexion deformity in addition, it is permissible to allow the muscle spasm to relax under a general anaesthetic so that the patient can be placed on a Jones abduction frame.

Old-standing fixed adduction deformity can be corrected by a subtrochanteric osteotomy.



FIG 93—Flexion contracture of a tuberculous knee, being corrected in a bent Thomas knee bed splint Note band supporting the head of the tibia.

Correction of flexion contracture of the knee

(ii) *Knee*.—Flexion contracture can be overcome slowly on a Thomas knee bed splint. The splint is bent to fit the contraction, the below-knee extension tapes are kept taut, and the splint is gradually straightened every other day (Fig. 93). Care is taken to support the head of the tibia to avoid subluxation of the knee. When the knee is to be arthrodesed, a little flexion is an advantage because, when the joint surfaces have been excised and the knee has been extended, the tight soft tissues at the posterior aspect of the joint allow a close and fairly stable fit of the bones, which aids the union.

Correction of deformities

(iii) *Ankle and foot*.—Equinus, varus and valgus deformities can be corrected by the repeated application of plasters.

follows the excision appears to do no harm, and is not aggravated by movements of the shoulder.

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(i) *Hip.*—Traction in the line of the deformity usually overcomes rigid *adduction deformity. When there is a pronounced flexion deformity in addi*tion, it is permissible to allow the muscle spasm to relax under a general anaesthetic so that the patient can be placed on a Jones abduction frame.

Old-standing fixed adduction deformity can be corrected by a subtrochanteric osteotomy.



Fig. 93.—Flexion contracture of a tuberculous knee, being corrected in a bent Thomas knee bed splint. Note band supporting the head of the tibia.

Correction of flexion contracture of the knee

(ii) *Knee.*—Flexion contracture can be overcome slowly on a Thomas knee bed splint. The splint is bent to fit the contraction, the below-knee extension tapes are kept taut, and the splint is gradually straightened every other day (Fig. 93). Care is taken to support the head of the tibia to avoid subluxation of the knee. When the knee is to be arthrodesed, a little flexion is an advantage because, when the joint surfaces have been excised and the knee has been extended, the tight soft tissues at the posterior aspect of the joint allow a close and fairly stable fit of the bones, which aids the union.

Correction of deformities of ankle and

(iii) *Ankle and foot.*—Equinus, varus and valgus deformities can be corrected by the repeated application of plasters

(iv) *Wrist.*—Palmar flexion deformity can usually be corrected by the repeated application of plasters. In old-standing disease the joint can be excised.

(6) Treatment of systemic complications

Importance of combined management in complicated cases

When other systems are involved, such as the respiratory and genito-urinary tracts, modification of the treatment of the joint is frequently necessary. In the presence of pulmonary tuberculosis, an artificial pneumothorax can be induced with the patient on a frame or other splint. The general open-air treatment is equally beneficial and when the skeletal tuberculosis is quiescent operative measures such as nephrectomy can be carried out. Fortunately, treatment of the other systems (apart from relaxation therapy for the lungs) can be delayed until the lesion of the joint is quiescent, and every open-air hospital should have a number of beds for such cases in which the patient can have the benefit of the combined treatment and supervision.

9. INDICATIONS FOR SURGICAL INTERVENTION

Operation in the quiescent phase

Tuberculous joints should be operated on only if the disease is quiescent. Attempts at fusion during the acute stage are dangerous, even though the fusion is extra-articular.

In children arthrodesis of joints should rarely be done (Jones, 1908 and 1909), with the possible exception of the hip, and more rarely the knee. From about the age of 12 years, successful ischio-femoral arthrodesis of the hip has been performed; it allows freedom from splintage. As regards the knee, a local fusion can sometimes be carried out, preserving the epiphyseal plates, and allowing the limb to continue its growth.

Operation has no bearing on the time factor

It should be impressed on patients that operative treatment does not in any way limit the duration of open-air treatment. It acts as a safeguard against recurrence of disease, and the substitution of internal splintage for external Nor does it obviate the need for the maintenance of general health at the highest level for the rest of life.

10. OPERATIVE TECHNIQUES

The best end-result

The best end-result of a tuberculous joint is a bony ankylosis, because such a result is safe and will stand any amount of wear and tear.

The old-fashioned method of excising a joint, though it may be successful in the knee and the sterno-clavicular joint, is useless for the hip, and extra-articular bone grafting is necessary.

Arthroplasty has little place in the treatment of tuberculosis of joints, with the possible exception of an old-standing arthritis of the elbow in which healing has occurred.

Failure in arthrodesis

The difficulty of securing a bony ankylosis in a tuberculous joint is nearly always due to the disease itself. Grafts are sometimes eroded and non-union occurs. Provided that the disease is thoroughly quiescent, the presence of an abscess does not necessarily mean failure. The abscess cavity can be excised. If an acute abscess is [illegible] tered, which can be distinguished by the appearance of the pus, the [illegible] should be abandoned temporarily.

Types of arthrodesis of the hip

[illegible] esis of [illegible] *moral a* [illegible] This useful [illegible]throdesis designed by Brittain [illegible] carri [illegible] e age of [illegible]. Disease of the hip joint

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(1) Arthrodesis of the hip

...s of ...rodesis ...e hip

(i) *Ischio-femoral arthrodesis.*—This useful arthrodesis designed by Brittain (1942) can be carried out from the age of 12 years. Disease of the hip joint

[illegible] from the [illegible]

performed, and a cortical tibial graft is placed across the osteotomy and into the ischium, using two osteotomes. The lower osteotome is removed and the graft is slid into place along the upper osteotome (Fig. 94). The shaft of the femur is displaced inwards, and a plaster spica is applied for many months. In a successful case the tibial graft hypertrophies and produces a firm strut uniting the femur to the pelvis, short-circuiting the hip, which recalcifies and heals (Fig. 95).



Fig. 94.—Shows the osteotome in position Another osteotome is slid in below it to open up the space, and a tibial graft is slid along into position.

Benefit of the ischio-femoral arthrodesis

The benefit of this operation, especially in children, is that freedom from splinting is allowed at a comparatively early date, and so knee movement rapidly returns and the lower limb develops normally. Following disease of the hip joint, there is all too often a grossly deformed knee and a short, withered limb.



Fig 95.—Same case as in Fig. 94, 12 months later Note hypertrophy of the graft, the hip itself has ankylosed by bone.

Failures are due to invasion of the graft by disease, fracture of the graft, failure to displace the femur inwards, non-union of graft to the cut surface of the femur and failure of the graft to engage the pelvis.

(ii) *Ilio-femoral arthrodesis.*—This is used when the disease spreads downwards into the ischium, or when an abscess appears under the neck of the femur, the calcification of which can be seen on a skiagram. A graft is taken either from the tibia or from the ilium, and inserted from the outer aspect of the ilium to the greater trochanter. If a tibial graft is used it can be driven upwards through a hole made in the trochanter and into the ilium The flat iliac graft is inserted into a slot cut in the ilium, femoral neck and trochanter. A plaster spica is applied for many months.

Failures are due to absorption and non-union of the graft, and to sinus formation.

(iii) *Intra-articular and extra-articular arthrodesis.*—This is a combination of ilio-femoral arthrodesis with excision of the joint.

(iv) *Osteotomy.*—In adults this procedure has given good results. A simple subtrochanteric osteotomy is performed and the shaft of the femur displaced inwards. A plaster spica is applied until the osteotomy has united. This has the affect of altering the line of weight-bearing from the hip to the pelvis.

The amount of abduction of the femoral shaft following an arthrodesis must be accurately worked out, so that the shortening is overcome as far as possible. This can best be done by careful clinical judgement at the time of operation or at the first change of the spica which should be done under a general anaesthetic.

(2) Arthrodesis of the knee

Excision and arthrodesis of the knee

Arthrodesis is carried out through a vertical or curved incision. Excision must include the synovial membrane, accessory ligaments and all the diseased tissue. The joint surfaces are removed with a saw or shaped with chisels and gouges, until a close fit is obtained. Cavities in the bones are cleared out. This procedure is frequently sufficient, and a plaster spica extending from the metatarsal necks to the pelvis is applied. When the disease is thoroughly quiescent and tuberculous pus is not encountered, internal fixation can be used, either by means of long stainless-steel nails or of bone grafts. The nails are inserted across the excision after the skin has been closed through separate stab wounds, and they are removed after a month.

Bone grafting, however, helps and accelerates the union. Many methods can be used. A graft may be fashioned from the tibia and slid upwards into a slot prepared for it in the femur. The cross-graft method is perhaps preferable to others because it gives such good internal fixation. Twin grafts are cut from the same or opposite tibia, and inserted in a crossed manner from below upwards through the tibial condyles into the femur. They are inserted along beds prepared for them with chisels. The cortex of the femur should be penetrated and a tight fit obtained, care being taken not to distract the bony [illegible] the other in the coronal [illegible] ell [illegible] nd bony union is usually complete in 3 months. If much caseous matter is encountered bone grafts should not be used, because they tend to be invaded by disease. The position of fusion should be just a few degrees of flexion.

Failures are due to recrudescence of the disease, distraction of the bony surfaces causing non-union and invasion of the grafts by disease.

(3) Arthrodesis of the ankle

Arthrodesis is carried out through an antero-lateral incision. The articular cartilage is excised until healthy bone is encountered. The soft tissue structures are very scanty and are excised. A graft is slid downwards from the tibia, across the joint into the talus. The foot is placed in a few degrees of equinus and held in plaster, usually for 4–6 months. For the first 2 months the plaster should extend from the metatarsal necks to the thigh. Then a below-knee walking plaster is applied until osseous union is complete.

Failures are due to the same causes as in the knee.

(4) Arthrodesis of the joints of the foot



Local operations on tarsal joints

(i) *Tarsal joints.*—When one tarsal joint alone is affected, an excision of the joint and a tibial graft placed across it is sometimes successful (Fig. 96).

(ii) *Metatarsals and phalanges.*—Excision and bone grafting of a metatarsal shaft have met with some success, but conservative treatment is usually enough (Fig. 97). Amputation of affected toes is the best treatment, and if the metatarso-phalangeal joint is involved, a portion of the metatarsal should be excised also.



Fig. 96.—Tuberculosis of the astragalo-scaphoid joint, treated by excision of the joint and tibial graft.





Fig 97 —Excision of almost all of the first metatarsal for tuberculosis, and replacement by a cortical tibial graft

(5) Arthrodesis of the shoulder

All adult cases should be fused in the optimal position. A combined intra-articular and extra-articular arthrodesis is usually necessary. The joint is exposed through a "sabre-cut" incision in the sagittal plane over the point of the shoulder, and the disease is cleared from the head of the humerus and the glenoid fossa. The diseased soft parts (scanty) are excised, and there is then very little bone left.

The spine of the scapula and the clavicle are divided and the acromion process is turned downwards into a slot prepared for it in the greater tuberosity (Fig. 98 (a) and (b)). This is not easy, because of the scanty bone and its poor quality. To [illegible]

difficulty another method of extra-articular grafting has been tried. A graft is placed across the axillary floor, through an incision along the posterior axillary wall. It is attached to the axillary border of the scapula and the shaft of the humerus (Brittain, 1942). Both methods are followed by plaster spica fixation for many months.

Failures by the first method are due to the frequency of fibrous ankylosis and non-union of acromion and great tuberosity. Failures by the second method are due to fracture of the graft, and failure of the graft to unite to the scapula or humerus.



(a)



(b)

FIG. 98.—(a) Immediate post-operative skiagram of intra-articular and extra-articular arthrodesis of the shoulder. Acromion shown embedded in greater tuberosity of the humerus. (b) Same case as (a) 12 months later. Note the strong bridge of bone uniting the acromion and the greater tuberosity.

(6) Arthrodesis of the elbow

Excision of the joint through a posterior vertical incision is usually sufficient, followed by plaster spica fixation until fusion occurs. Ankylosis may be hastened and made more certain if the bones are bridged by one or more tibial grafts, if there is sufficient bone to receive them. The usual position is just below a right angle with the hand pronated. A spica should be applied for the first 2 months. Then a plaster is applied from the knuckles to the axilla until fusion has taken place.

Failures are due to recrudescence of the disease and mechanical difficulties of fixation of bone of poor quality.

(7) Arthrodesis of the wrist

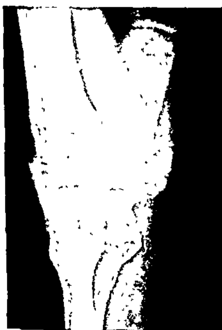
There are many methods of arthrodesis. All the diseased tissues should be excised; this may be difficult because of the complicated relations of the articulation. The carpo-metacarpal joints should be included to avoid the pain which sometimes follows more conservative procedures. A broad flat iliac graft, or a wide tibial osteo-periosteal graft, helps this extensive fusion. It is always possible to retain rotatory movements of the forearm by not

interfering with the inferior radio-ulnar joint or, if this joint is involved in the disease, by excision of the lower end of the ulna.

The wrist is exposed through a vertical incision and the tendons are retracted. All the diseased tissue is excised, as indicated above, and a groove made in the radius, the remains of the carpal bones, and the bases of the outer four metacarpals. The wrist is placed in 20° of dorsiflexion and the graft fitted into its bed. The wrist and elbow are immobilized in plaster of Paris, with the fingers free, until fusion has occurred. (Fig. 99 (a), (b) and (c). Failures are due to inadequate excision of the disease, and non-union or fracture of the graft.



(*a*)



(*b*)



(*c*)

FIG 99 —(*a*) Tuberculosis of the wrist Quiescent phase, bones are well calcified (*b*) and (*c*) Tuberculosis of the wrist; same case as in (*a*) Post-operative skiagrams to show the solid bony fusion following excision of the diseased part and osteo-periosteal grafting

Metacarpals.—These do not as a rule require surgery, because conservative treatment is adequate.

Phalanges.—In the presence of a sinus in an adult it is wisest to amputate the finger.

(8) Amputations

Amputation is sometimes necessary to save life. It is indicated when a patient, after a period of general treatment, is obviously going downhill, and the *When to amputate*

diseased joint is deteriorating, as seen by the rapid and continuous formation of pus. This applies to the knee, foot and wrist. The hip joint is a more formidable problem. The choice lies between disarticulation and the clearance of as much of the diseased pelvis as possible, or a wide excision of the joint (Bankart, 1933). In the upper extremity a major amputation is most often necessary for extensive disease of the wrist joint with multiple secondarily infected sinuses.

Amputation in tuberculosis of the foot

In children and young adults the disease in the foot tends to involve one bone and remain localized to it. Conservative treatment is usually successful. In adults the disease is apt to be much more widespread throughout the tarsal joints and if, after a period of conservative treatment, there is no sign of the disease limiting itself, then a below-knee amputation should be done without delay. In the elderly, amputation is the treatment of choice.

Multiple tuberculous lesions

When a patient is suffering from multiple lesions, complete extirpation of one or more by amputation will often determine recovery, by decreasing the strain on his recuperative powers. Thus, when a patient has a tuberculous knee and a tuberculous phalanx of a finger and toe, amputation of the finger and toe will allow the healing processes to concentrate on the knee.

In the presence of tuberculosis of other systems, when the patient is having an uphill struggle, amputation of a limb may turn the tide in his favour.

Amputation of the hand

One is naturally reluctant to amputate a hand even when there are severely infected sinuses in the wrist. The patient's general condition must be the guide. Amputation should be performed if he is losing ground.



FIG. 100.—Tuberculous hip with sinuses and large sequestrum in roof of acetabulum. This was cleared out by operation. The sinuses healed and then an ischio-femoral arthrodesis was carried out.

Osteomyelitis

(9) Sequestrectomies

When the bones have become secondarily infected, large sequestra form and must be removed before healing can be expected. Wide exposures of the joints may be necessary (Fig. 100).

Resettlement of the patient

(10) After-care

Special care must be taken to ensure that the patient shall return to suitable conditions of home life and employment. Every effort must be made to resettle the patient and to enlist the interest of his employers. He should have rest periods during the day, plenty of open air, and sleep with the window open, or, better still, sleep in a hut in the garden. He must be gradually restored to work, and if this is too heavy or unsuitable, he must be retrained. Many patients return successfully to wage earning and can support their families once again. The patient reports to his tuberculosis authorities,

and to the hospital in which he was treated, for a clinical review and radiographic examination. When a joint has been successfully fused no external splintage is necessary. When a weight-relieving caliper or a below-knee double steel for ankle or foot has been applied, then these and other splints will require supervision, usually from the hospital in which he had his initial treatment.

Unfortunately, relapses do occur, and may be due to the lesion being insufficiently stabilized, or to unsuitable conditions of living. Severe intercurrent illness, such as influenza, may also cause a relapse.

11. RESULTS OF TREATMENT

No large statistical follow-up can be given but, generally speaking, if the patient has one joint alone affected and is placed in the suitable surroundings indicated above, then a favourable result can be anticipated. With more than one joint involved, and especially if other systems are affected, chronic invalidism is a frequent result.

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KIDNEY AND URETER—CYSTS

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PART I
CYSTS OF PARASITIC ORIGIN

1. HYDATID DISEASE

(1) Aetiology

208.] For a general account of the aetiology and pathology the reader is referred to the section on Hydatid Disease, p. 46. As in the case of other tissues and organs which can be infected only by the systemic blood stream, kidney infection is rare because the larva has to travel first of all through the liver, heart and lungs.

(2) Pathology

Tissue reaction

The rate of growth of the embryo is very slow, and the cyst may not give rise to symptoms for as long as 20 years. As it acts as a foreign body to the tissues, a reactionary wall of fibrous tissue forms around it. The cyst itself consists of two layers, an outer membrane and an inner or germinal layer. From the latter grow small papillae or buds which eventually become small cysts or brood capsules. Inside them appear the scolices or new taenia heads. On the other hand, the mother cysts may develop daughter cysts identical in formation and in their method of propagating brood capsules or scolices.

Primary, secondary or invasion cysts

A cyst of the kidney derived directly from the embryo is termed *primary*. If formed from a scolex, piece of detached germinal layer, brood capsule or daughter cyst which has migrated from some other part of the host, it is called *secondary*; but if the kidney has been penetrated from an infected adjacent organ it is known as an *invasion cyst*. The usual site of implantation is the renal cortex, and the anterior surfaces of the upper and lower poles. It may also arise in the loose tissue of the sinus and be adherent to the renal pelvis.

(3) Symptomatology

"Closed" or "open" cyst

[illegible] depends upon whether the cyst remains intact [illegible] ruptures (when it is known as an

The closed cyst does not give rise to any symptoms until it is large enough to press upon adjacent structures. Until then the patient is unaware of its existence and is perfectly well.

When the cyst begins to leak, renal colic occurs, followed by difficulty in passing water or even by retention of urine, due to an accumulation of membrane in the bladder. The urine may contain pieces of this membrane, daughter cysts, hooklets and scolices. A common manifestation of the rupture of a cyst is urticaria. At a later date kidney and bladder become infected with the *Bacillus coli* and other organisms, when the predominant symptoms will be those associated with pyelonephritis and cystitis—fever, dull aching pain in the loin and suprapubic region, scalding and frequency of micturition.

Urticaria



FIG 101.—Hydatid cyst of right kidney (open variety). The contents comprise numerous daughter cysts. There is a secondary cyst of the upper pole. (*From the museum of St. Peter's Hospital for Stone and Other Urinary Diseases, London.*)

(4) Diagnosis

(i) *Closed variety.*—A correct diagnosis can be made only when there is a palpable tumour in the loin, and pyelography demonstrates that a calyx is blunted or that there are areas of calcification (Figs. 102 and 103).

(ii) *Open variety.*—In the first instance the diagnosis is that of an infection of the urinary tract, but skiagrams and laboratory tests reveal that hydatid disease is the primary lesion.

(iii) *Urine.*—In the open variety the urine contains hooklets, scolices, daughter cysts or pieces of membrane. The hooklets are pointed at one end and barbed.

(iv) *Serum test or intradermal reaction of Casoni.*—This test consists in the injection of 0 1–0 3 cubic centimetre of sterile hydatid fluid into the skin. If the test is positive a weal surrounded by a zone of erythema appears in 10–20 minutes, and is followed some hours later by the appearance of a larger area of congestion.

Casoni reaction

(v) *Examination of operation material.*—Hooklets will be found in the hydatid cyst fluid. The laminated membrane round the cyst has a characteristic histological structure.

(5) Complications

The cyst may press upon the renal pelvis and cause the formation of a hydronephrosis. If the latter becomes infected a pyonephrosis results.

Accumulation of membrane in the bladder will cause chronic retention. Urethritis follows from the repeated passage of hooklets and scolices.

(6) Treatment

Nephrectomy

Provided that the other kidney is healthy, the only treatment is nephrectomy. The prognosis is good. During the removal of the organ care must be taken

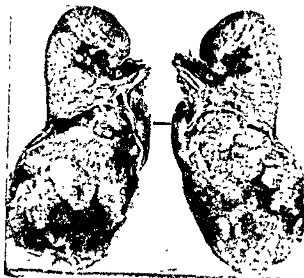


FIG. 102.—Hydatid cyst of left kidney (closed variety). Removal by operation from a woman aged 48 years. The wall of the cyst is calcified (*see* pyelogram, Fig. 103) and its contents are gelatinous. (*From the museum of St. Peter's Hospital for Stone and Other Urinary Diseases, London.*)



FIG 103.—An ascending pyelogram of the left kidney in a woman aged 48 years, showing a calcified hydatid cyst of the lower pole (closed variety). (*From the museum of St. Peter's Hospital for Stone and Other Urinary Diseases, London.*)

not to rupture the cyst: such a catastrophe will lead to infection of the perinephric tissues.

2. SCHISTOSOMA HAEMATOBIUM

A detailed account of this disease is unnecessary here as it very rarely appears in the form of cysts in the renal pelvis.

Examination of urine

The diagnosis is made by examining the last few drops of urine passed, or the centrifugal deposit from them, under a $\frac{2}{3}$-inch or $\frac{1}{6}$-inch objective. The ova with their terminal spines are easily recognized.

Irritation of the skin is an initial symptom, followed by an irregular temperature and the appearance of a vesicular rash. Later haematuria occurs, due to invasion of the bladder. Treatment consists in the intravenous administration of antimony sodium tartrate. The disease can be cured by this drug. A total of 30 grains must be given, and the course should be intensive in order to prevent the parasite from acquiring a resistance to antimony.

PART II
CONGENITAL CYSTS

1. POLYCYSTIC DISEASE

(1) Aetiology

This disease makes its appearance from birth to late middle life, but symptoms occur usually between the ages of 40 and 60 years. If observed in the newly born, death probably occurs in under a month, but when signs and symptoms do not manifest themselves until adult life, the patient is likely to survive until the sixth decade. The disease is more common in females, and is liable to occur in families. As a rule both kidneys are involved, but the disease is *more extensive on one side.*

(2) Pathology

The surface of the kidney is studded with masses of cysts ranging in size from that of a pin's head to that of a grape; the thin walls of these cysts are semi-transparent, of a clear brown, yellow or blue colour, and spread throughout the substance of the kidney.

The organ may reach enormous proportions, weighing as much as 15 pounds. The cysts contain a clear brownish or yellow fluid, which comprises all the principal constituents of normal urine with occasional red blood cells, albumin and oxalate crystals.

The tissue between the cysts is fibrous, with compressed glomeruli and tubules. Other organs besides the kidney may be cystic, generally the liver or pancreas.

(3) Symptoms

In the infantile form the large size of the infant's abdomen is noticeable at birth, and the tumour is easily felt on palpation. In a few days, or under a month, death takes place from uraemia

The course of the symptoms in the adult may be followed, according to Thomson-Walker (1914), through three periods.

(a) Latent period.
(b) Period of renal tumour with symptoms.
(c) Period of renal failure.

(a) Latent period

During the latent period the disease is not discovered until the kidney has reached a large size. It is then that the patient draws the doctor's attention to the presence of a swelling in the loin. The tumour is painless and moves on respiration. At this stage the urine is normal and renal function tests show no evidence of failure. A careful examination of the opposite loin may show this kidney to be larger than normal, and palpation of the anterior surface of the liver may reveal the presence of nodules The latter is not a constant manifestation, but when it is present there is no doubt left in the mind of the clinician as to the nature of the tumour in the loin

(b) Period of renal tumour with symptoms

This period commences with the development of symptoms. The tumour is now so large that it is causing some distress. It has the position and contour of a renal tumour and in spite of its size is movable. To the palpating hand it conveys a sense of elasticity If the patient is thin the cysts on the surface of the kidney can be felt. There may be a dull ache in the loin. In 40 per cent of cases the first indication is haematuria; it can be so profuse that it causes ureteric colic. Polyuria is common, the urine containing a small quantity of albumin with a low specific gravity. It is unusual to find casts.

(c) Period of renal failure

The stage of renal failure occurs, as a rule, between the ages of 50 and 60. The urine gradually becomes more and more scanty and symptoms of uraemia develop. The patient complains of headache and loss of appetite. The face is pale and the tongue brown and dry. There is often vomiting and also diarrhoea. Sometimes convulsions occur and finally the patient dies in coma.

Rise in blood-urea

During this period the urea in the blood steadily rises and towards the end may reach 300–500 milligrams per cent. The urine is loaded with albumin.

(4) Diagnosis

(a) Clinical

Skiagrams and cystoscopy

Diagnosis is not difficult if a tumour can be felt in each loin, but when the swelling is unilateral it is impossible to determine its nature accurately without the aid of skiagrams and possibly of cystoscopy.

A large hydronephrosis will give rise to similar signs, and if haematuria is present a diagnosis of new growth cannot be excluded.

(b) Radiography

Intravenous pyelography presents a characteristic picture which is unlike that of any other renal lesion. The shadow of the pelvis is smaller than normal. The calyces appear as long spindle-like structures with their cup-shaped ends rather larger than normal. The large size of the kidney is readily demonstrated.

(c) Cystoscopy

When haematuria is present cystoscopy is indicated to exclude the presence of a bladder tumour. One or both effluxes will be blood-stained.

(5) Prognosis

The disease is always fatal but life may extend over many years during which time the patient enjoys good health. Death from uraemia occurs between the fifth and sixth decades, but there are cases on record in which the terminal illness has not appeared until 70 years of age (Jossand; Morson, 1938).

(6) Treatment

As polycystic disease is incurable the treatment is concerned with the symptoms. The period of the patient's survival is measured by the amount of active renal tissue, and therefore nephrectomy, both partial and total, is contra-indicated.

Nephrectomy contra-indicated

Puncture of the cysts (Rovsing's operation) is sometimes performed as an attempt to relieve the pressure upon healthy tissue, but at best the operation is a palliative procedure, for in due course the cysts again become distended.

The technique consists in exposing the kidney through an oblique lumbar incision and puncturing the cysts on the anterior and posterior surfaces, so far as wide retraction of the wound permits. As the kidney diminishes in size so it is possible to deliver more and more of it out of the wound, when further cysts can be punctured. The contents should be evacuated with an aspirating needle, taking care to avoid damaging healthy renal tissue. The kidney is replaced in the loin, dusted with sulphathiazole and penicillin powder, and the wound closed without drainage. If haematuria is severe it is treated by blood transfusion and rest in bed; morphine is given for the relief of colic. When signs of uraemia appear, saline and glucose must be given by continuous drip and the patient must be persuaded to drink large quantities of fluid. A temporary improvement often follows this treatment, but the patient, owing to the absence of healthy renal tissue in sufficient quantity to maintain life, gradually relapses into coma.

Sulphathiazole and penicillin

2. DERMOID CYSTS

Dermoid cysts of the kidney are very rare. They contain hair, sebaceous material and degenerative products

(1) Symptoms

There are no marked symptoms in the early stages, but should the cyst enlarge there will be backache and loin tenderness.

(2) Diagnosis

The tumour is seldom palpable and cannot be diagnosed without the aid of an x-ray examination. Pyelography may show a pressure effect upon one or more calyces and areas of calcification.

Pyelography

(3) Treatment

In some cases it may be possible to remove the cyst without undue interference with the renal parenchyma, but if the whole of its wall cannot be excised nephrectomy is indicated.

3. SOLITARY CYSTS

Since evidence favours the view that solitary cysts are acquired, rather than congenital in origin, they are considered in Part III.

PART III
ACQUIRED CYSTS

1. RETENTION CYSTS

(1) Aetiology

These cysts are multiple and rarely larger than a marble. They occur in chronic nephritis.

(2) Pathology

The pathology is that of chronic interstitial nephritis. The cysts are of surgical interest only when unilateral and associated with hyperpiesis.

(3) Symptomatology and diagnosis

Persistent headache, high blood-pressure and polyuria are the common indications of a commencing chronic nephritis.

The diagnosis of these cases is made by the physician and, when medical treatment has failed to relieve symptoms, he is inclined to refer the patient to the surgeon in the hope that he can effect a cure.

and possibly casts in the urine coming from the affected side will reveal this. Urine from the opposite side will be normal. Intravenous pyelography may show no excretion of dye from the diseased organ. (In some cases excretion

ization and one specimen only is found to contain abnormal constituents, ascending pyelography should be performed in order to avoid submitting the patient to instrumentation on two separate occasions.

(4) Treatment

The surgeon, having satisfied himself that only one kidney is affected, should perform nephrectomy on the diseased organ.

(5) Prognosis

The immediate results are satisfactory. A number of cases are now on record in which a permanent cure of hyperpiesis can be legitimately claimed.

2. SOLITARY CYSTS

(1) Aetiology

There is a divergence of opinion as to their origin; while some workers claim that they are congenital, others think they are acquired after inflammatory occlusion of one or more tubules. The evidence favours the latter view, for they are invariably found in those of advanced years. The condition is rare, more frequent in women and usually unilateral.



Fig. 104.—A solitary cyst of the left kidney. (*From the Archway Museum of the L.C.C.*)

(2) Pathology

The typical form is a thin-walled transparent cyst projecting beyond the surface of the kidney (Fig. 104).

It is often as large as a child's head and appears to develop from the renal cortex into which the wall is merged. On the kidney aspect the renal tissue is depressed to form a cup-shaped cavity. The cyst wall consists of fibrous tissue. Very occasionally there may be more than one cyst.

(3) Contents

A clear yellowish fluid, containing albumin and salts, fills the cyst. Haemorrhage into the cyst is not uncommon.

(4) Symptomatology

These cysts do not give rise to any symptoms until haemorrhage occurs into them or until they become large enough to press upon adjacent structures. A sudden haemorrhage causes the patient to complain of sharp pain, associated with nausea and vomiting; pressure causes a dull aching pain in the loin. If the patient is thin, a tumour connected with the kidney can be felt in the loin.

(5) Diagnosis

The tumour may be mistaken for a hydronephrosis, but pyelography will determine its nature. The pyelogram of a solitary cyst shows compression of one or more calyces; the outline of the pelvis, contrary to that in hydronephrosis, is normal. Renal function tests do not show any evidence of uraemia. The urine may contain a trace of albumin but no other abnormal constituents.

(6) Treatment

Sutures contra-indicated

The loin must be explored by operation, when the cyst will be found protruding from one or other pole of the kidney. It is easily shelled out by making an incision at the junction of its wall with renal tissue and separating it from the latter with a blunt dissector. Bleeding is infinitesimal. A cup-shaped cavity is left, the wall of which consists of atrophied renal tissue. There is no need to close this space by sutures, for if they are used a haematoma may form, which will eventually become a haemorrhagic cyst.

(7) Prognosis

Immediate results are satisfactory, but when chronic inflammatory changes are present in the kidney a guarded prognosis must be made. Should the opposite kidney become affected at a later date, there will be deterioration in the patient's general health.

3. HYDROCALYCOSIS

(1) Aetiology

Outlet obstruction

A cyst may arise from the dilatation of a solitary renal calyx due to obstruction of its outlet. This obstruction may be inflammatory in origin or due to a stone. The difference between this type of cyst and the solitary or serous variety is that there is a communication in the former with the renal pelvis. The opening may be no larger than will admit a piece of horsehair.

(2) Pathology

The hydrocalyx, unlike the solitary cyst, does not present itself beyond the renal cortex, and may be overlooked unless the kidney is carefully palpated. The condition must not be mistaken for those d[illegible]e to mechanical obstruction at th[illegible]ract. The c[illegible]

(3) S[illegible]

Th[illegible]ually haematuria accompanied by dull aching pain in the loin. The urine contains pus and red blood cells, and when cultured there is a growth of *B. coli*

(4) Diagnosis

This can only be made by a process of exclusion, but pyelography will sometimes reveal the narrow communication between the calyx and the cyst cavity. It may also demonstrate the shadow of a stone impacted in the outlet of the calyx.

(5) Treatment

The kidney must be explored and the diagnosis established by palpation and by needling the cortex. A round-bodied needle of large size inserted into the substance of the kidney will readily demonstrate the presence of the cyst cavity. Nephrectomy is the correct procedure, for attempts to save the organ by drainage of the cyst will only lead to a secondary infection and the formation of a urinary fistula.

Needling

Nephrectomy

(6) Prognosis

If the disease is unilateral the condition will be cured by nephrectomy.

PART IV
MEGALO-URETER

1. PHYSIOLOGICAL

During pregnancy the whole of the upper urinary tract undergoes hypertrophy, and dilatation of the calyces, pelvis and ureter occurs. The latter may be distorted to a considerable size. This condition is of surgical interest only if the investigator has to distinguish between a pathological and physiological cause for the dilatation as shown by pyelography and ureterography. The presence of albumin in the urine, or of urinary symptoms, calls for x-ray examination of the kidneys, ureters and bladder; only an experienced urologist

can determine to what extent the variations of the ureter from the normal may be deemed pathological in origin. The previous history of urinary symptoms will establish a correct diagnosis, but the cases of greatest difficulty are those in which a symptomless megalo-ureter of some years' duration manifests itself in pregnancy. Only a complete investigation by cystoscopy, ureteral instrumentation and radiography will ascertain whether or not dilatation of the duct is due to a pathological lesion.

2. INFLAMMATORY

(1) Aetiology

Ureterectasis

Repeated attacks of inflammation of the mucous membrane of the ureter will lead to its dilatation. If the oedema is chronic then the overstretching of the wall causes permanent ureterectasis. The concertina-like action of the renal pelvis which occurs in intermittent obstruction of the ureter due to inflammation is similarly observed in the latter. The ureterectasis disappears when the inflammation subsides (Morson and Graham, 1940).

(2) Pathology

B coli inflammation

The most common cause is the inflammation produced by the *B. coli*. The dilatation involves not only the ureter but also the pelvis and calyces. Other organisms which produce this condition are the *Staphylococcus aureus* and the tubercle bacillus. The tissue changes are the same as those which result from the obstruction of any duct—hypertrophy and dilatation, followed by atony.

(3) Symptoms

During the acute stage there is ureteric colic and haematuria. Should the condition become chronic there is a persisting infection of the upper urinary tract, giving rise to an ever-present pyuria and frequency of micturition.

(4) Diagnosis

The whole urinary tract must be investigated by the aid of urine analysis, cystoscopy, ureteral instrumentation and ureterography. It is then possible to exclude the other causes of megalo-ureter such as stricture, stone and the idiopathic variety.

If a stricture or stone is present the ureteral catheter will be held up at the point of obstruction, and a ureterogram will show its position. In inflammation the catheter passes without difficulty along the whole duct. The dilatation in the idiopathic variety is very much greater and is confined to the ureter.

(5) Treatment

Chemotherapy

Treatment is that of the infection which can be controlled by the sulphonamide drugs and penicillin. Surgical operation is not indicated.

3. OBSTRUCTIVE

(1) Aetiology

There are three types of obstruction which may produce megalo-ureter.

(a) Stricture at the vesical end of the ureter.

(b) Calculus.

(c) Aberrant vessel.

(2) Pathology

Congenital stricture is a fairly common cause; the narrowing is limited to the intravesical portion of the ureter. Calculi readily lodge in this part of the duct; if they are large they become impacted and give rise to dilatation of the ureter. Very occasionally an aberrant vessel will arise from the aorta, cross the middle portion of the ureter and enter the lower pole of the kidney. This condition is associated with abnormal arrangements of other blood-vessels. Above the artery the ureter is dilated to a considerable size (Fig. 105).

(3) Symptoms

The mechanical obstruction of the ureter causes stasis of urine in the kidney and the ureter above the site of the lesion. There is a dull ache in the loin on the affected side, varying in intensity according to the quantity of fluid intake by the mouth. At times there may be an attack of colic. The patient is always conscious of "something being not quite right" on the side of the abdomen in which the lesion is present.

Pyonephrosis

In course of time infection occurs and the symptoms are intensified. A late result may be the formation of a pyonephrosis.

Before infection occurs the urine will contain a few red blood cells and some pus cells. It will be sterile on culture.



FIG. 105—A retrograde pyelogram showing on the left side a dilatation of the upper half of the ureter. At operation an aberrant vessel was found arising from the aorta. This artery crossed the ureter at right angles in its middle third and then passed up to the lower pole of the left kidney. It thus formed a loop. The megalo-ureter was cured by division of the artery. (*A. W. Badenoch's case.*)

(4) Treatment

Stricture.—Under a general or local anaesthetic, ureteral bougies are passed through the vesical part of the ureter with the aid of a cystoscope. The dilatation must be continued until the bougie is so firmly gripped at the ureteral orifice that the mucous membrane is in danger of being torn.

Division of mucous membrane

A month later this instrumentation should be repeated. Further treatment is indicated only if there is a recurrence of symptoms. Should the stricture be impassable an attempt may be made, with special scissors passed through an operating cystoscope of the Joly or Winsbury-White pattern, to divide the mucous membrane of the ureteral orifice. This is a very difficult manoeuvre and seldom successful. The tissues divided must include the muscle coat. Bougies can then be passed until the stricture is fully dilated.

Suprapubic cystotomy

If all attempts at instrumentation fail, a suprapubic cystotomy must be performed with the patient in the Trendelenburg position. With the aid of Morson's electrically-lit retractors the base of the bladder is visualized. The ureteral orifice is divided by a narrow-pointed scalpel on a long handle, after which the dilatation is completed by the passage of ureteral bougies.

Aberrant vessel.—Intravenous pyelography demonstrates the position of the aberrant vessel as it crosses the ureter (Fig 105).

The obstruction is always in the upper half of the ureter. The vessel can be exposed and divided by the usual kidney incision in the loin.

Should the ureter below the obstruction fail to distend after division of the artery, an incision should be made in the dilated part and bougies passed down the tube into the bladder. Thus the narrowed portion of the ureter is stretched. Complete recovery may be expected.

4. NON-OBSTRUCTIVE OR IDIOPATHIC

(1) Aetiology

This variety of megalo-ureter is often associated with poliomyelitis, spina bifida and cerebrospinal syphilis. If none of these lesions can account for the condition it is attributed to an error in mesenchymal development, or some failure in neuro-muscular co-ordination. It may exist undetected for many years.

(2) Pathology

Association with other congenital abnormalities

The disease may affect one or both ureters, and should it continue in adult life may result in degeneration of the kidney. It is frequently associated with other congenital abnormalities, some of which are extra-urinary, such as Hirschsprung's disease.

Infection of the upper urinary tract is an early complication, owing to stasis of urine.

(3) Symptoms

There is abdominal discomfort and a feeling of distension, but in childhood the first manifestation is the presence of pus and red blood cells in the urine.

(4) Diagnosis

As with the other varieties of megalo-ureter the whole urinary tract must be investigated. Cystoscopy reveals no abnormality of the ureteric orifices, but the urinary effluxes are sluggish and there is an absence of muscular movement. Instead of being expelled with some force the urine tends to trickle out of the ureteric orifice.

(5) Treatment

Various operations have been devised to relieve this condition, and the fact that several have been tried indicates that none is really satisfactory. Dilatation of the ureteric orifice is ineffective, but transplantation of the ureter into the upper part of the bladder is helpful because it gives drainage to the stagnant urine. Operations upon the nervous system consist of presacral neurectomy and spinal anaesthesia. Occasionally a good result is obtained by dividing the presacral nerve in the hollow of the sacrum. This is a simple operation and consists of dividing the peritoneum over the promontory of the sacrum, when the nerve, which is sometimes double, is seen lying on the bone. It is then traced down to the hollow of the sacrum and about 3 [illegible] of it excised.

As the injection of Stovaine (amylocain[illegible] ride) into t[illegible] has been effective in a number of cases o[illegible] g's disea[illegible] it has been thought that it might pr[illegible] satisfac[illegible]

idiopathic megalo-ureter, but this has not proved to be so; temporary improvement has been reported in a few cases, however.

(6) Prognosis

If the disease is unilateral and all palliative means have failed to relieve symptoms, nephrectomy and ureterectomy will effect a cure.

Renal failure Bilateral megalo-ureter of idiopathic origin is a fatal disease and the patient eventually succumbs to renal failure due to infective pyelonephritis.

PART V
URETEROCELE

(1) Aetiology

Ureterocele is a prolapse of the ureter and occurs in two varieties. In one there is a prolapse of the whole thickness of the ureteral wall and in the other of the mucous membrane alone. It is an uncommon lesion, females being more prone to it than males. A number of cases of it have been recorded in girls between 2 and 5 years of age. It is sometimes bilateral.

(2) Pathology

The tumour is a globular cyst attached by a narrower base in the region of the ureteric orifice. It may be as large as a golf ball or as small as a pea. It consists of a double layer of mucous membrane; bladder mucosa externally, and ureteral mucosa internally. On some part of the surface the ureterio orifice can be seen. This is small and frequently stenosed. Occasionally calculi are present in the cavity. *Calculi* In the female the cyst is prone to prolapse into the urethra and appears at the external meatus as a plum-coloured swelling. In this position it may become strangulated and undergo necrosis.

(3) Symptoms

In the early stages the symptoms are those of a lesion of the upper urinary tract, namely, lumbar pain referred along the course of the ureter. Infection being often present, there may be attacks of fever with frequency of micturition.

When the tumour has reached a large size the symptoms are similar to those of a stone in the bladder—frequency of micturition, haematuria and pain at the end of the urethra. In the female there may be sudden retention of urine, due to the prolapse of the cyst into the urethra.

(4) Diagnosis

Diagnosis is made with the cystoscope. In the region of the ureteric orifice a pink, semi-translucent tumour is seen, covered by bladder mucous membrane. The ureteric orifice is on the summit of the swelling as a rule, if it is on

the under aspect it is difficult to find. While under observation the cyst may vary in size and even collapse like a pricked bubble. Should it present itself at the external meatus it can, if not strangulated, be reduced into the bladder.

(5) Treatment

Cystoscopic diathermy

The tumour can be destroyed by cystoscopic diathermy. If it is prolapsed into the urethra the patient should be given a general anaesthetic and placed in the lithotomy position, when the cyst is seized with tissue forceps, pulled out as far as possible, and the pedicle divided with the endothermy current.

The prognosis is satisfactory and a cure can be anticipated.

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— and Graham, W. H. (1940) *Brit J. Surg*, 27, 540
Thomson-Walker, J. W. (1914) *Genito-Urinary Surgery*. London; Cassell.
[References to other titles are given under Kidney and Ureter—Cysts, in the Index Volume.]

KIDNEY AND URETER—DENERVATION OF THE KIDNEY

By J. B. OLDHAM, V.R.D., F.R.C.S.
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1. ANATOMY

209.] The kidney receives its nerve supply through the renal plexus which *Renal plexus* arises from several roots—from the aortico-renal ganglion, the semilunar

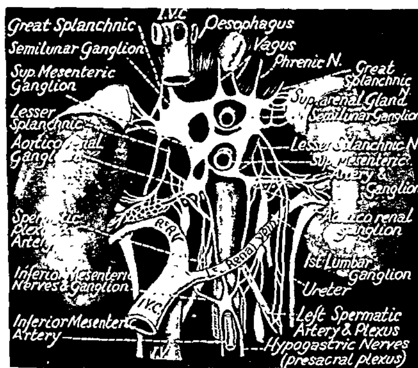


Fig. 106—Anatomy of the renal nerves The inferior vena cava has been divided and the peripheral end has been turned down.

ganglion, direct from the lesser splanchnic nerve and from the least splanchnic nerve if it is present At their origin these roots are widely separated; they

Periarterial formation

converge upon the renal artery and [illegible]

plexus (Fig. 106).

Relation to other nerves

Through its roots the plexus is in close relationship with the nerve supply to other organs. In addition, three important nerves arise from the plexus or its roots: (1) the spermatic or ovarian nerve, (2) the inferior mesenteric nerve which provides the main branch going to the presacral nerve and also supplies the left half of the colon, and (3) the nerves to the upper ureter arising from the plexus close to the hilum.

Site for denervation

Over the distal third of the renal artery, before it divides into its secondary branches, the nerves constituting the plexus are concentrated into a compact network, and here it is easy to perform a complete denervation of the kidney without injury to the nerve supply of other organs.

2. PHYSIOLOGY

Absence of secretory fibres

The modern theory of urinary secretion does not invoke the necessity, or admit the existence, of secretory fibres to the kidney ; secretion continues indefinitely after all connexions between the central nervous system and the kidney have been severed.

Vasomotor fibres

Ring muscle system

Sensory fibres

The principal function of the renal nerves is vasomotor. In addition there are motor fibres controlling the musculature of the organ itself. There is a series of ring muscles (Fig. 107) encircling the bases of the papillae, the neck of the calyces and the pelvi-ureteric junction; these are controlled by the renal nerves. The sensory fibres running in the plexus supply mainly the papillae, calyces and pelvis. (*See* Table.)

TABLE

NERVES	STIMULATION	DEPRESSION OR SECTION
Nerves to the blood-vessels	Vaso-constriction ↓ Decreased flow of blood through kidney ↓ Anuria or oliguria	Vaso-dilatation ↓ Increased flow of blood through kidney ↓ Increased urine of low specific gravity
Nerves to the pelvis, calyces and papillae	Contraction of sphincters ↓ Increased intra-pelvic pressure ↓	Relaxation of sphincters
Sensory nerves	Pain	Anaesthesia of kidney

3. INDICATIONS FOR DENERVATION

Theoretical indications

If the concept of the physiology of the renal plexus outlined in the previous paragraph is accepted, then denervation might be expected to give favourable results in any renal condition in which pain demands relief, the sphincters exhibit hypertonus, an increased blood supply to the kidney is desirable or diuresis is indicated.

Practical indications

In practice the operation is indicated only in renal pain

of unknown origin, in non-mechanical hydronephrosis, or for pain due to nephroptosis.

(1) Nephralgia

Not infrequently we meet with patients who complain of pain which is undoubtedly renal in origin, and yet the most careful investigation fails to reveal any appreciable abnormality in the kidney or its function. Whatever may be the source of this pain which, lacking a definite pathology, is sometimes called "nephralgia", whether it is a true neuralgia or neuritis or is a sympatheticotonus causing increased intrapelvic pressure, division of the renal nerves is as logical a treatment as is division of the fifth nerve for trigeminal neuralgia. The work of French urologists, the brilliant surgery of Hess in America and of Harris in Australia and the results reported by the writer and others in Great Britain, all give practical proof of the value of renal denervation in this type of case.

Sympatheticotonus

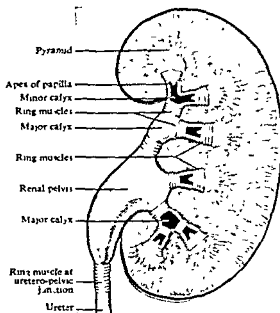


Fig. 107.—Diagrammatic representation of the ring muscle system. The anterior walls of the minor calyces and lower major calyx have been cut away to expose the tips of the papillae.

(2) Non-mechanical hydronephrosis

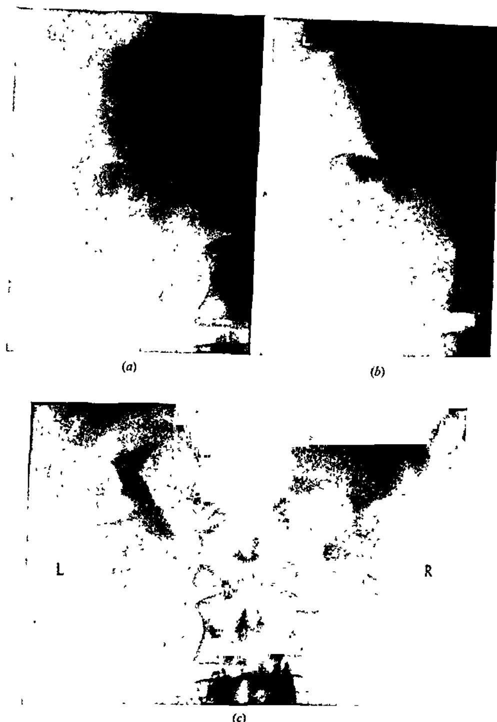
There is an increasing belief that non-mechanical hydronephrosis is due to inhibition of the normal relaxation of the sphincters of the calyces and pelvis and is analogous to Hirschsprung's disease. It is recognized that section of the sympathetic nerves to the colon will cure Hirschsprung's disease in so far as it will relieve the stasis and produce regular evacuation, but the diminution in the size of the dilated colon may be both slight and slow. The cases reported in the literature, and personal experience, all prove that in non-mechanical hydronephrosis the pain can be cured, the pelvic stasis relieved, the renal function improved and the size of the pelvis gradually diminished by denervation alone. (Figs. 108 and 109.)

Similarity to Hirschsprung's disease

(3) Nephropexy

Renal denervation is suggested as a substitute for nephropexy as it is believed that the pain and other symptoms in nephroptosis are due to nervous reflexes and not to mechanical obstruction of the ureter or to pressure on the viscera. The results of nephropexy are notoriously uncertain, and it is possible that some of the successful cases are due to an unwitting denervation being done while the kidney is being fixed.

Nephropexy uncertain cure



(a) (b)

(c)

Fig. 108.—Female, aged 26 years; domestic servant. Three years' recurrent attacks of L. renal colic. Forced to give up work. In 1932, denervation of L. kidney. No pain since operation. Working regularly.

(a) Before operation. Skiagram taken 30 minutes after injection of Uroselectan B. Marked hydronephrosis; poor concentration of dye.

(b) Two years after denervation. Skiagram taken 30 minutes after injection of Uroselectan B Hydronephrosis slightly diminished; improved concentration of dye.

(c) Seven years after operation. Skiagram taken 30 minutes after injection of Uroselectan B Hydronephrosis has disappeared.



(a)



(b)

FIG. 109 (*a*), (*b*), (*c*) and (*d*).—Female, aged 15 years; mill hand Ten months' recurrent attacks of L. renal colic, lasting for 3 days with intervals of a week. Incapacitated for work. In 1933, denervation of L. kidney. No pain since operation.

(*a*) Before operation. Skiagram taken 10 minutes after injection of Uroselectan B Hydronephrosis of left kidney, defective secretion of dye.

(*b*) One year after denervation of left kidney Skiagram taken 10 minutes after injection of Uroselectan B Hydronephrosis diminished; marked improvement in secretion of dye.



(c)



(d)

FIG 109.—(*cont.*)

(*c*) Before operation. Skiagram taken 30 minutes after injection of Uroselectan B. Large left hydronephrosis.

(*d*) One year after operation. Skiagram 30 minutes after injection of Uroselectan B. Hydronephrosis now very slight.

Contra-indications

Operation is called for only in a very few selected cases of nephroptosis *All cases of general visceroptosis should be excluded. Operation should be* avoided in confirmed neurasthenics and in cases in which neither pyeloscopy

nor pyelography shows evidence of pelvic stasis. In all cases prolonged conservative treatment must be tried before surgery is considered.

4. INVESTIGATION AND SELECTION OF CASES

Urological investigation

A thorough investigation must be made in every case and, if necessary, repeated again and again. The investigations must include chemical and bacteriological examination of the blood, and of the urine from each kidney; cystoscopy with injection of indigo-carmine; and intravenous and retrograde pyelography. When retrograde pyelography is being done the fluid is injected slowly until the patient begins to complain of pain in his side, and he is then asked if this pain is like the pain he usually has. Patients seldom have any hesitation in answering with a definite "Yes" or "No", and only if this test is positive should denervation be considered.

"Pain reproduction" test

When the "pain reproduction" test and other criteria are satisfied, it should be explained to the patient that, although the kidney is causing his pain, it is not seriously diseased and that he should try conservative treatment for a short time. If he returns complaining that he is no better, the urological investigations should be repeated and only if they confirm the original findings is denervation advised. The final decision is only made when at operation the diagnosis is confirmed and there is no indication for any other form of treatment.

Conservative treatment

5. OPERATION

The operation requires great gentleness and patience. Good exposure and lighting of the renal pedicle are essential, otherwise the operation is difficult and dangerous.

Anaesthesia

Unilateral spinal anaesthesia, using light Nupercaine, with the addition of gas and oxygen, gives complete muscular relaxation and with it the peritoneum falls away from the incision, giving perfect exposure of the renal pedicle.

Instruments

No special instruments are needed, but the operation is certainly easier if the surgeon has very long and fine dissecting forceps, haemostats and scissors, especially if these are curved or angulated.

Exposure

The kidney is exposed as in any other renal operation. An oblique incision with sub-periosteal excision of the twelfth or eleventh rib (Mouat, 1939) is particularly suitable.

Stripping the pedicle

The fatty tissues are stripped away to expose the renal artery and vein. The easiest and most natural way to clean the adventitious tissue of the vessels is to work from the kidney inwards towards the aorta, but this method is prone to cause venous haemorrhage—the only real danger of the operation. The adventitious tissue and nerve plexuses invest the renal vessels and their branches like a glove, and must be removed by working outwards from the inner end of the pedicle towards the kidney.

It is impossible to give definite directions as to where the stripping and denervation of the pedicle should be started—it depends upon the length of the pedicle, the relationship of the artery and the vein and other considerations—but usually it is easiest to start on the back of the pedicle as the renal vein overlaps the artery in front.

Removing the nerve fibres

When the pedicle has been cleared of fatty tissue the nerve plexus will be seen running along the renal artery and its branches. The fibres of the plexus are picked up over the inner end of the artery on a grooved director (Fig. 110) or on a strabismus hook and divided with scissors. The cut distal ends of the nerves are picked up with fine forceps and gently stripped towards the kidney where they are divided again. This process is repeated until the artery and its main branches are stripped quite clean for at least one inch.

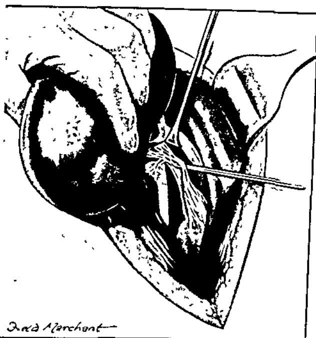


FIG 110.—Fibres of plexus picked up on grooved director prior to division.

There is no need to strip the nerves right up to the hilum. That it is unnecessary to do so has been shown when discussing the anatomy of the plexus; it is also unwise, for it is difficult to strip the fibres right up to the kidney without being troubled by venous bleeding.

If an aberrant artery is present, the nerve plexus accompanying it must be excised.

Denervation of upper ureter

After the pedicle has been denervated the upper ureter is separated from its bed and its fascial sheath; no attempt is made to strip it as clean as the renal artery. Finally the kidney will be attached solely by its denervated pedicle and ureter and separated from any connexion with its nerve supply.

Carbolic acid

When the artery has been stripped as clean as possible, it is painted over with 10 per cent carbolic acid which will destroy sympathetic fibres (De Luca, *1931*) *without damaging the renal vessels or ureter. Quite apart from* any effect it may have in destroying any fine nerve fibres which have not been cut, the acid rapidly whitens them so that they can be recognized and excised.

I have avoided combining renal denervation with any other operation on the kidney—nephropexy, decapsulation or plastic operation on the pelvis—not because there is any objection to this, but because I wished to obtain unequivocal evidence of the effects of denervation.

6. RESULTS OF DENERVATION

Secretion of urine

Most patients complain of severe pain for 2 or 3 days after operation, but this has all gone within a week. For a day or two the secretion from the denervated kidney is markedly diminished, but then it rapidly increases until it is much more than normal. After denervation indigo-carmine appears sooner in the

urine, the jets from the ureteric orifice are more frequent and the volume of the stream is greater, but the concentration of the dye is not as deep as in the urine from the normal kidney. All these changes diminish in 3-6 months and there is then little to choose between the secretion from the normal and the denervated kidney.

Following denervation it is impossible to cause discomfort or pain, no matter how much the pelvis is distended. Between 6 and 12 months after operation some sensation returns, but a marked hypoaesthesia remains always; even after 5 years, over-distension of the pelvis only causes a slight ache.

Provided that the renal arteries or veins are not damaged, the renal function is in no way harmed by denervation. This has been proved by careful follow-up and investigation of patients who have either had both their kidneys, or their only remaining kidney, denervated.

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KIDNEY AND URETER—GROWTHS

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1. DEFINITION

210.] The term, growth, is used here to include neoplasms of the kidney, renal pelvis and ureter.

2. AETIOLOGY

Workers in aniline dyes are subject to papillary tumours of the kidney as well as of the bladder; squamous-celled carcinoma is sometimes associated with stone and often with long-standing infection. A maldeveloped or ectopic kidney is more prone to new growths than is a normal kidney. No age group is immune but malignant tumours occur most frequently before the age of 5 years or after that of 40 years. Males are affected more often than are females.

3. SURGICAL ANATOMY

Surgical approach

The retroperitoneal position of the kidney facilitates surgical approach from the lumbar region, and the close relationship to the colon, duodenum and

diaphragm makes this route generally desirable. A large tumour will project into the peritoneal cavity and may be more readily removed by a transperitoneal operation.

4. PATHOLOGY

Most renal tumours are malignant, benign tumours forming less than 7 per cent of the total. The classification of renal tumours may be given as follows:

I. KIDNEY

	Benign	*Malignant*
Epithelial	Adenoma	Adenocarcinoma (granular celled) Hypernephroma (clear celled)
Connective tissue	Fibroma Lipoma Myoma Angioma	Sarcoma (spindle celled)
Mixed	Adeno-fibroma	Wilms's tumour (children) Fibrosarcoma with tubules (adults)

II. RENAL PELVIS AND URETER

Benign	Malignant
Papilloma	Carcinoma Transitional celled Squamous celled

(1) Benign tumours

Benign tumours are relatively unimportant; adenomas are small single encapsulated tumours sometimes associated with chronic nephritis. Small fibromas are frequent post-mortem findings. An angioma is a tumour of blood-vessels which grows at the apex of a pyramid; it may cause severe haematuria. An adeno-fibroma may reach a large size, one weighing 22 pounds having been removed by Gordon-Taylor (1930), but most large tumours prove to be malignant. Benign papillomas of the pelvis or ureter spread widely in the urinary mucous membrane and tend to become malignant if they are left untreated.

(2) Malignant tumours

Adenocarcinoma

Adenocarcinoma of the granular-celled type is the rare form of malignant epithelial new growth. It starts in the renal tubules and may be diffuse, tubular or papillary in histological structure. Macroscopically, the growth is more uniform than is the clear-celled type, but it is rather more invasive and tends to spread by the lymphatics (Fig. 111).

Hypernephroma

Clear-celled carcinoma was given the name, hypernephroma, by Grawitz (1883) who considered that it arose in an adrenal rest. It is the commonest renal tumour, accounting for 70 per cent of the total. Arising beneath the capsule at the upper or lower pole, it may reach a considerable size before being discovered. The frequent appearance of fibrous encapsulation is due to compression of the surrounding renal tissue. The tumour contains fat and has a yellowish-red colour with patches of necrosis and haemorrhage, and intersecting fibrous strands.

Microscopically it consists of large clear vacuolated cells containing globules



FIG 111.—Adenocarcinoma. The branching papillary processes are composed of a vascular core covered by a layer of columnar cells with granular cystoplasm. (× 140.) (*By courtesy of Dr. A. C. Thackray.*)

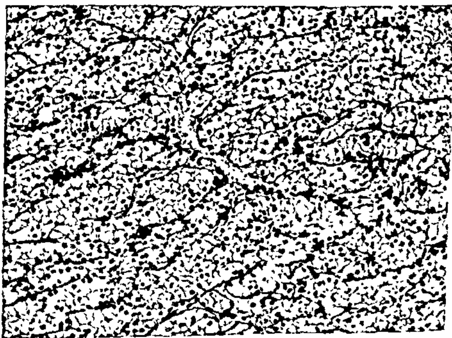


FIG. 112.—Hypernephroma. Section shows a tumour made up of columns and groups of the typical clear cells, each with a round, darkly-staining nucleus. (× 140.) (*By courtesy of Dr. A. C. Thackray.*)

of fat, arranged in rows or masses according to the plane of section (Fig. 112).

It disseminates by the venous blood stream, causing metastases in the lungs, liver, bones and opposite kidney. Lymphatic invasion also occurs, as well as direct local infiltration. The presence of a metastatic deposit may be the first indication of the disease.

Sarcoma

Sarcoma is uncommon. It may occur at any age and may attain a large size. It arises in the capsule or in perivascular connective tissue, and is usually of the spindle-celled variety. It is highly malignant.

Wilms's tumour

Of the tumours of mixed origin, Wilms's tumour—the embryonic adeno-myosarcoma—affects children from birth to the age of 5 years; rare cases have been reported in adults. It arises anywhere in the kidney and forms a large tumour with a dirty-white or brown colour on section. It may appear to be encapsulated and is sometimes bilateral. Microscopically it is seen to consist of a basis of immature connective tissue with muscle fibres, fat, glandular epithelium, cartilage and occasionally bone or teeth. The varying proportions of its constituent tissues account for the variations in its nomenclature. Spread is by the blood stream, and growth both of the primary tumour and of metastases is rapid.

Fibrosarcoma

There is an unusual type of fibrosarcoma containing epithelial tubules, which occurs rarely in adults.

Squamous-celled tumours

Malignant tumours of the pelvis or ureter may be squamous-celled or papillary (transitional). The squamous-celled tumours are uncommon; they tend to occur in more elderly subjects, and are often associated with chronic infection or with stone. They form firm nodular masses which infiltrate the kidney, and their spread is largely lymphatic, although blood-borne metastases occur.

Papillary tumours

Papillary tumours resemble those of the bladder in that their malignancy varies from the benign papilloma to the infiltrating carcinoma. They are not so uncommon as was formerly supposed; Macalpine (1947) has reported a personal series of 19 cases.

These tumours appear as papillary masses on the pelvic mucosa and may be large and exuberant or may form a low mossy carpet. In the course of time the pedicles become wide and confluent and infiltration takes place. Sometimes a single calyx is involved, but in late cases the whole pelvis and ureter, as well as much of the bladder, may be covered with growth. This is their main method of spread, by implantation, or by multiple foci of origin, but lymphatic and vascular dissemination can occur.

Ureteric growths

Tumours of the ureter are likely to be secondary to tumours of the renal pelvis, but primary growths do occur. Because of the small size of the ureter and the lack of any great amount of ensheathing fat or connective tissue, all malignant ureteric growths become adherent at an early stage, and are often inoperable because of local fixity.

Metastases

Although any tumour of the kidney will gradually destroy that organ, death is usually due to metastases. All the malignant renal tumours can spread by the blood stream; the greatest incidence of metastases is in the lung, in the liver and in the bones. Of the bones affected the spine is most frequently involved, accounting for 46 per cent of the total (Graham 1947) followed in order of frequency by the ribs, pelvis and humerus.

5. CLINICAL PICTURE

Haematuria

The main clinical manifestations of a renal tumour are haematuria, swelling and pain. Of these haematuria is the most constant. It occurs at some stage in the majority of cases and is the initial symptom in 80 per cent. The bleeding is often profuse, blood being intimately mixed with the urine; when it is very severe there may be clots, but worm-like ureteric clots are rare. Micturition is painless, except for the passage of clots. Haemorrhage is intermittent; it may cease spontaneously after a few hours or days.

The onset of bleeding is determined by invasion of the pelvis by the tumour and thus, in part, by its site in the kidney. A tumour at a pole has to grow larger than one in the centre before reaching the pelvis, and the initial sign in such a case is more likely to be swelling than haematuria. This occurs in about 60 per cent of cases, a fact which indicates the late stage reached before advice is sought. Swelling is the first clinical sign in most cases of Wilms's tumour, haematuria being late. Such tumours grow rapidly and there may be a visible swelling when the child is first seen.

Swelling

The swelling occupies the loin and is palpable bimanually; at first it is mobile and moves on respiration, but later it becomes fixed by direct local spread. It is partly dull and partly resonant from overlying colon, but a large tumour is usually quite dull and pushes the colon downwards below it.

Pain

Pain is not an outstanding feature, although it is present at some stage in the majority of cases. It is of a dull aching character and renal colic occurs only if clots are passing. The combination of painless haematuria with a vague pain in the loin is very suggestive of renal new growth, especially if the haematuria starts before the pain.

Other clinical features are sometimes noted; there may be some increased frequency of micturition, due to irritation from blood clot; rarely, the bleeding is so rapid as to produce clot retention in the bladder.

Increased frequency of micturition

Persistent slight albuminuria or microscopic haematuria are of common occurrence.

Varicocele

Varicocele, especially when occurring on the right side after the age of 35, is a significant finding, since idiopathic varicocele is rare on this side. It is due to pressure of the growth or to engorgement of the capsular veins which anastomose with the spermatic veins. It does not disappear on recumbency but does so after nephrectomy and it is not a contra-indication to operation.

Slight pyrexia is present in a few cases, and is said to be due to necrosis and absorption of toxins or to a reaction to a vascular metastasis.

Hypertension

Hypertension is occasionally noted but may be incidental, and the general effects of any malignant disease, such as wasting and anaemia, are late, but are all too commonly seen when the patient first seeks advice.

Enlargement of the liver or evidence of pulmonary or osseous metastases are sometimes the first clinical signs, and a pulsating swelling in a bone should always arouse suspicion. In a case recorded by Bland-Sutton (1922) the bony swelling preceded evidence of the renal growth by 6 years.

6. SPECIAL AIDS TO DIAGNOSIS

Early diagnosis is essential in a tumour which metastasizes so readily, and it is possible only if the patient and his doctors are prepared to follow any suspicious symptom to the logical conclusion of full investigation.

The two special investigations which are diagnostic in the greatest number of cases are cystoscopy and retrograde pyelography. Other forms of examination which may be helpful in a patient in whom the possibility of renal neoplasm exists are radiography and examination of the urine for albumin, blood and cells. A plain x-ray picture in a well-prepared patient may show an alteration in the renal outline, or calcification in a hypernephroma.

Urine examination and radiography

(1) Intravenous pyelography

Intravenous pyelography is a useful preliminary to retrograde pyelography; it may give a positive diagnosis, and it will give evidence of the function of the other kidney.

Perirenal oxygen injection has been used to make the radiological renal outline more definite; its use in the diagnosis of neoplasm will be limited to a very few difficult cases, and it may then be combined with pyelography.

Perirenal injection of oxygen

(2) Cystoscopy

Cystoscopy must be performed in every case of haematuria more especially when it is of the "painless" type, that is, not accompanied by symptoms of acute cystitis. The strict observance of this rule would facilitate the earlier diagnosis of both bladder and kidney tumours. Cystoscopy should be performed during haematuria. If there is a growth in the bladder it may be difficult to obtain a sufficiently clear medium, but if there is a renal tumour this is the golden opportunity for finding the source of the bleeding. So much may depend upon the result of cystoscopy that it should be undertaken with extreme care and an opinion should not be given until the medium has been thoroughly cleared. If the patient is bleeding when he is first seen, it is advisable to undertake cystoscopy at once under a local anaesthetic, using a small examination cystoscope. If a blood-stained efflux is seen from one ureteric orifice, the other must also be examined; the case may be one of bilateral polycystic disease. If bleeding has stopped, cystoscopy may be delayed. The passage of a ureteric catheter to determine the side of bleeding is sometimes misleading, but it may be

Time for cystoscopy



FIG 113 —Hypernephroma, right kidney. Compression of the upper calyx and dilatation of the lower The crescentic filling defect is due to a growth in the upper pole



FIG. 114.—Solitary cyst, left kidney. The crescentic filling defect is due to a large cyst between the upper and lower poles.

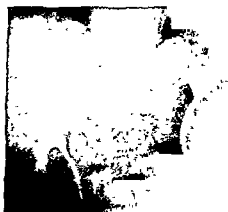


FIG. 115.—Papillary carcinoma, left renal pelvis. The lower part of the pelvis is occupied by a villous tumour which shows variations in density.



diagnostic in the case of a ureteric growth. The catheter is obstructed and bleeding occurs when attempts are made to advance it.

Ureteric growth

A low ureteric growth may project from the ureteric orifice and a piece can sometimes be taken for biopsy.

(3) Retrograde pyelography

When cystoscopy is delayed until intravenous pyelography has been performed, preparations can be made for retrograde pyelography to be

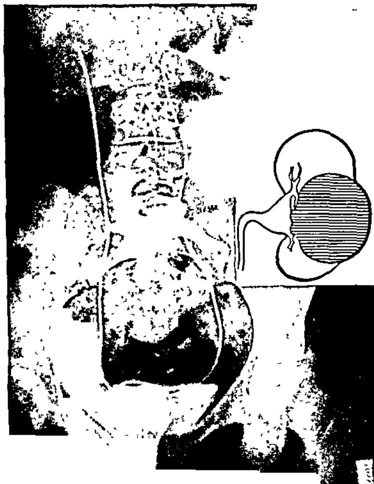


FIG 116—Hypernephroma, left kidney. The calyces are compressed *and the pelvis is narrowed.*

performed at the same time as cystoscopy, and a different form of anaesthesia may be desirable. The technique of pyelography does not differ from that used for other conditions; it is advisable to catheterize both ureters in order that a double pyelogram can be made if it is necessary to compare the two sides. The pelvis must be well filled, and it is therefore better to use a dilute solution of one of the intravenous pyelographic media (for example, Uroselectan B) rather than sodium iodide, because some degree of overfilling is not then of such importance.

Pyelographic appearance

The interpretation of the pyelogram may be difficult; the changes produced vary with the type and site of the growth. The commoner changes are as follows:

Compression of calyces

(i) *Compression or obliteration of calyces.*—The compression sometimes produces a characteristic crescentic filling defect in a calyx (Fig. 113). Such



FIG. 117.—Hypernephroma and hydronephrosis, right kidney The growth in the lower pole caused extrinsic pressure on the ureter.

a defect can be caused by any rounded structure compressing a calyx, and is seen in polycystic disease and in solitary cyst (Fig. 114).

Pelvic filling defect

(ii) *Encroachment on the lumen of the pelvis.*—This will be caused earlier by a pelvic growth (Fig. 115), but cortical growths will also produce it (Fig. 116).

Dilatation

(iii) *Secondary pyelectasis*—Some degree of dilatation may occur even in the absence of obstructive changes at the pelvic outlet (Fig. 117). With a growth of the ureter or at the pelvi-ureteric junction a hydronephrosis may obscure the signs of growth (Fig. 118).



FIG. 118.—Papillary carcinoma, left pelvi-ureteric junction. The growth caused intrinsic obstruction and hydronephrosis.



FIG 119.—Hypernephroma, left kidney. Both pelves are dilated but the left calyces are narrowed and elongated, giving a spider-leg deformity. Bleeding was observed from the left ureter and the diagnosis was confirmed at operation

Spider-leg deformity

(iv) *Elongation of the calyces* (Fig. 119)—This is the so-called spider-leg deformity; the calyces are long and thin, in contrast to the appearance of the polycystic kidney in which they are lengthened and dilated. The change



FIG 120.—Hypernephroma, right kidney. The calyces are elongated and the interval between the upper and middle calyces is increased by the tumour.

appears to be produced by a tumour growing in between two calyces and the angle between them is altered at the same time (Fig. 120).

A similar change is sometimes caused by fibrosis.

(v) *Displacement.*—Displacement of the ureter, or of the kidney itself, may be caused by the growth. This change can be produced by extrinsic pressure from a retroperitoneal tumour (Fig. 121), but in this case the regularity of the pelvic outline is not affected.



FIG. 121.—Retroperitoneal sarcoma distorting the left ureter and displacing the kidney outwards.



FIG. 122.—Primary carcinoma of the ureter. There is an irregular filling defect with dilatation of the ureter and pelvis above it.

(vi) *Irregularity.*—In the case of a papillary tumour of the pelvis the filling defect may show variations in density which are suggestive of the medium passing in between the fronds of a growth (Fig. 115). This appearance, however, can also be given by blood clot in the pelvis.

(vii) *Ureteric growths.*—With a growth in the ureter pyelography may not be possible, but a ureterogram will show a filling defect. This will be localized in the case of a primary carcinoma, with dilatation above if the obstruction can be passed (Fig. 122). In diffuse papillomatosis the ureter is generally dilated and the ureterogram shows mottled areas of varying density; in many cases a picture cannot be obtained owing to difficulty in catheterizing the ureter.

7. DIFFERENTIAL DIAGNOSIS

A swelling in the loin should be recognized as renal or otherwise from its clinical features. Palpable bimanually, it moves downwards and inwards on respiration (before local fixation has occurred) and may show bands of resonance.

Spleen

Enlargement of the spleen may simulate a renal swelling but a notch should be palpable and it is everywhere dull to percussion. Tumours of the adrenal gland push the kidney downwards, and retroperitoneal tumours displace it outwards (Fig. 121). Colonic growths near the flexures may cause difficulty; they are resonant at first, and become fixed earlier than renal growths. Mesenteric cysts are usually movable from side to side.

Growths and cysts

Cause of renal enlargement

Diagnosis has to be made of the cause of renal enlargement; the conditions which most nearly resemble new growth are hydronephrosis, in which the swelling is smooth and reaches a large size without local fixity; polycystic disease, in which the swelling is nodular; and solitary cyst, in which the differentiation may be impossible although haematuria is less common. Renal calculi may also cause enlargement.

The source of haematuria must first be localized to the upper or lower urinary tract. This is done by cystoscopy, and the differentiation of causes of renal haematuria is made by pyelography. The importance of these two investigations must again be stressed, and the pyelogram, as a rule, is decisive.

8. PROGNOSIS

The outlook, as judged by published figures, must be regarded as poor, the 3-year survival rate after operation varying from 19 per cent (Garceau, 1909) to 30 per cent (Mintz, 1938); a recent series gives 28 per cent (Graham, 1947). There is some evidence that granular-celled adenocarcinoma is more lethal than is hypernephroma (Muir and Goldsmith, 1935).

vis

The pelvic growths are less likely to spread by the blood stream, but they have a greater tendency to local dissemination within the urinary tract. The general outlook is rather better than that for renal cortical tumours, Joly (1933) reporting a 27 per cent 5-year survival rate.

ter

In primary tumours of the ureter it is rare for patients to survive for more

metastases. There is much variability in the rate of growth of renal tumours and some of them give rise to late metastases, even after 15 years. A solitary metastasis does not preclude surgical treatment, and even after gross invasion of the renal vein we have seen a 5-year survival.

Wilms's tumour

The tumour with the worst reputation is the Wilms's tumour of children; there are reports of isolated cases of survival but in the majority of patients secondary deposits develop early in the lungs, liver or opposite kidney. The effects of the recent trend towards pre-operative irradiation cannot yet be assessed.

9. CHOICE OF TREATMENT

Every renal tumour must be regarded as malignant; even the simple papilloma can destroy life. Treatment must therefore be early and radical. The methods available are surgery and irradiation, either singly or in combination.

(1) Irradiation

The radio-sensitivity of renal tumours varies, just as does their malignancy. In general the more malignant tumours are the more radio-sensitive, and Wilms's tumour often shows a striking regression after irradiation although the more differentiated adult tumours are little affected.

On the other hand the metastases of hypernephroma may be very radio-sensitive, but the beneficial effect is usually only temporary. It is doubtful whether any cure has been obtained by irradiation alone, and its chief value is in supplementing surgical treatment. Deep x-ray therapy is the most convenient form of application; radium needles may sometimes be used for isolated metastases.

It remains a matter for individual decision whether or not x-ray treatment should be given before operation. It may reduce the size of a large tumour and make nephrectomy possible, but it has a debilitating effect, it delays operation for about 2 months and it may predispose to wound disruption. The writer's practice is to use irradiation in Wilms's tumour, for which any method likely to improve the prognosis is justifiable, but not to employ it in adults except in special circumstances.

Post-operative irradiation should be given if there has been any local spread of growth beyond the kidney, or if glands in the hilum are invaded. It will not prevent vascular metastases.

For recurrences after operation x-ray treatment will often produce dramatic regression, but this is usually followed by the appearance of another more radio-resistant recurrence in a short period.

(2) Surgical methods

(a) Nephrectomy

The most satisfactory treatment is surgical extirpation of the diseased kidney. It is usual to remove the perinephric fascia also when dealing with a new growth, but this makes little difference to the outcome. It is important to minimize manipulation of the kidney during its removal, and to tie the vascular pedicle as early as possible. These factors may influence the surgeon, so that he chooses the abdominal rather than the lumbar route, but in our experience the advantages are theoretical rather than practical, and we prefer the lumbar route except in very large tumours.

(b) Nephro-ureterectomy

It is necessary to remove the whole of the ureter when dealing with a papillary tumour of the kidney or with a ureteric growth. It has been the practice to include a small disc of bladder with the lower end of the ureter, but Macalpine (1947) has drawn attention to the danger of opening the urinary tract, with the consequent possibility of spilling urine containing tumour cells. In such an event a local recurrence is rapid and inevitable. It is advised instead that the ureter be divided between clamps at its extreme lower end, using diathermy. The distal clamp is used to coagulate the vesical end of the ureter before ligation; the intramural part can be further coagulated by *cystoscopic diathermy*.

Whenever possible, the need for removal of the ureter must be determined by pyelography before operation, since it is not always easy to recognize the exact nature of the tumour at operative exposure. A papillary tumour is not so hard and it arises in the pelvis. In the early stage it feels rather like a piece of wet cotton-wool inside the pelvis. If the ureter is not removed and the tumour is found subsequently to be of the papillary type, the remainder of the ureter should be removed in a second operation 2 or 3 weeks later.

Secondary ureterectomy

Primary new growths of the ureter should be treated by nephro-ureterectomy; when the function of the other kidney is satisfactory there is no place for partial excision of the ureter and anastomosis.

10. PRE-OPERATIVE MANAGEMENT

Before operation it is essential to be certain of the presence of a functioning kidney on the other side; the preliminary investigations will have decided this Urgent nephrectomy for new growth is rarely needed but may be necessary for continued bleeding.

Renal function tests

The most useful tests of renal function are the estimation of blood urea, the indigo-carmine excretion test and intravenous pyelography.

Anaemia due to prolonged bleeding should be treated, according to its degree, by the administration of iron or by blood transfusion. Fluid intake should be maintained at a high level. Apart from treating any gross dietary deficiency or cardiovascular impairment, little time should be spent in pre-operative measures; the sooner a kidney growth is removed the better.

11. OPERATIVE TECHNIQUE

(1) Lumbar nephrectomy

Under adequate anaesthesia the patient is placed on his sound side, the lower leg being flexed with the upper leg straight, and the loin is opened out by the bridge or other means.

Incision

The incision runs from the renal angle obliquely downwards and forwards to a point above the anterior superior spine of the ilium. Adequate exposure is essential; posteriorly it can be increased by division of the external arcuate ligament (lateral lumbo-costal arch) or by section of the twelfth rib. In front it is limited only by the size of the abdomen, and in an obese patient there is room for a longer forward extension of the incision.

The diagnosis is confirmed by the finding of large veins in the perinephric fat, and there will be some inevitable bleeding from them. The kidney must be

turned forward and the pedicle secured as early as possible; mass ligation *Pedicle* is better than efforts to secure the vessels separately. Growth may be present in the renal vein, and care must be exercised not to dislodge it. Extension into the vena cava may be removable and, if other conditions are satisfactory, this step is justifiable. The ureter *should be followed down and divided;* it should then be cauterized, and ligated at a convenient level.

(2) Nephro-ureterectomy

In papillary or ureteric tumours the ureter is not divided but the incision is closed around it, leaving the kidney outside. The patient is turned on to his back and the lower end of the ureter is exposed through an oblique inguinal or paramedian incision. Division of the ureter should be performed by diathermy at the level of the bladder, the lower ureteric stump being well coagulated before ligation.

The cavity left after removal of the kidney is sprayed with sulphanilamide powder. Drainage is usually provided, but is not always necessary if haemo- *Drainage* stasis is complete.

The difficulties encountered are due to size, fixity and extension of growth, and the *operation may not be easy, especially on the right side, owing to the* proximity of the inferior vena cava, which may be involved. Arrangements for blood transfusion should always be made beforehand. *Blood transfusion*

(3) Abdominal nephrectomy

If the tumour is very large it may be removed by a transperitoneal route. The abdomen is opened through a rectus incision, extending from the costal margin to below the umbilicus; if further room is needed a right-angled extension can be made laterally from its centre. The peritoneum is divided at the outer border of the colon, which is reflected inwards; on the right side the duodenum is also mobilized. The renal pedicle is found by palpation; it enters high and runs obliquely downwards and outwards. It should be adequately exposed and secured by a double ligature, a blunt boomerang *Ligation of* needle or an aneurysm needle being useful for this purpose. A clamp is also *pedicle* applied on the proximal side before the pedicle is divided, and a ligature on the renal side prevents escape of blood which may contain tumour cells. The kidney is then mobilized from below and behind and is removed together with the perinephric tissue. The ureter is divided as low as is conveniently possible (except when dealing with a papillary tumour when it should be followed down to the bladder) A stab drain is placed through the posterior abdominal wall, and the peritoneal surface is restored posteriorly before closure of the anterior wound.

Difficulties arising during operation

Sudden bleeding from the pedicle or from the vena cava is at first alarming, *Haemorrhage* but it can be controlled by temporary gauze packing; after an interval the packing is removed piecemeal and the bleeding point can usually be secured. It is rarely necessary to leave forceps or packing in place, for removal after 72 hours. The use of a muscle graft, sutured over a rent in the inferior vena cava, *Injury to vena cava* is sometimes useful.

[illegible] *to*

Injuries to peritoneum and intestine

Any tear in the peritoneum should be sutured before the end of the operation.

The duodenum or colon may be injured when adhesions are dense; if recognized, the injury should be repaired at once. A fistula occurring later can usually be closed by the application of continuous suction and the administration of sulphasuccidine, but operative closure may be necessary.

12. POST-OPERATIVE CARE

Position

On his return to bed, the patient is placed on his back and inclined towards the side of operation. On recovery from the anaesthetic he is propped up in a sitting position in bed and thereafter he should be moved frequently to either side. It is a good plan to get him out of bed as early as circumstances permit, and this may be done on about the fourth day or sooner in many cases. Early ambulation prevents pulmonary, vascular and intestinal complications.

Sedatives

Opiates or other sedatives may be given freely; pain is often considerable after nephrectomy.

Fluids

After modern anaesthetics, fluids can be given by mouth in adequate quantity to make the routine use of intravenous fluids unnecessary, but any suggestion of renal failure may demand their use; blood-chemistry tests are of value in this respect.

Any considerable blood loss should be replaced by blood transfusion during or after operation.

Infection

Infection is not usually a prominent feature after nephrectomy, but if there has been soiling of the wound or much perinephric inflammation it is wise to give sulphonamides by mouth (for example, sulphadiazine, $\frac{1}{2}$ gramme, 6-hourly) as a prophylactic measure, and penicillin (100,000 units, 6-hourly) for the same purpose, reserving more frequent dosage for real need so as not to disturb sleep.

Drainage

The drainage tube, if one is used, can be removed on the second day if there is no infection or haemorrhage.

Flatulence and constipation are often troublesome owing to the proximity of the colon. Liquid paraffin, $\frac{1}{2}$ ounce thrice daily, should be started on the day after operation and an aperient given on the second evening. Enemas are usually necessary, but the greatest preventive of intestinal stasis is early ambulation.

Cardiac and pulmonary complications must be combated as they arise.

Follow-up

The later after-care demands regular examination with a view to the recognition of metastases, and skiagrams of the chest or suspected bones should not be omitted In the case of pelvic papillary tumours cystoscopy is a most important part of the follow-up.

The possession of only one kidney necessitates care in the avoidance of chills and urinary infection

13. RESULTS OF TREATMENT

The somewhat depressing figures quoted under the heading of Prognosis should not be a deterrent to active surgical or radiological treatment; in a case which at first appears hopeless, the patient may live for several years; in many of the cases diagnosed early, complete cures are achieved. Nevertheless, new growths of the kidney form one of the most serious types of the disease and efforts should be directed mainly towards their earlier diagnosis.

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[References to other titles are given under Kidneys and Ureters—Growths, in the Index Volume. The subject is also dealt with under the heading of Kidney, Surgical Diseases in the *British Encyclopaedia of Medical Practice* (1938), Vol. 7, p. 380.]

KIDNEY AND URETER—HYDRONEPHROSIS AND PYONEPHROSIS

By W. E. UNDERWOOD, O.B.E., F.R.C.S.
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PART I
HYDRONEPHROSIS

1. DEFINITION

211.] Hydronephrosis is essentially an enlargement of the renal pelvis and calyces, containing sterile urine. As a transient phase, infection may be present, but should this persist a pyonephrosis may follow.

2. AETIOLOGY

Obstruction

The condition has generally been ascribed to obstruction, but recent consideration has shown that this is not invariably the case. In pregnancy, for example, the enlargement may be demonstrated by intravenous pyelography before mechanical pressure by the foetal head can be responsible, and as the

condition disappears after parturition, hormone activity is now considered to be the cause. *Hormone activity*

In congenital hydronephrosis, although the antecedent conditions are present at birth, the enlargement has not yet occurred; the term, congenital, is therefore misleading. As this type is due to neuro-muscular dysfunction of the renal pelvis, dynamic is a more accurate term. *Congenital type*

In other cases hydrostatic back-pressure is the cause. There may be mechanical obstruction of the upper or occasionally of the lower urinary passages. This obstruction may be permanent and incomplete; but intermittent complete obstruction can have the same effect. *Mechanical type*

The condition may be unilateral or bilateral, the latter form generally being secondary to some abnormality in the lower urinary tract; this must be considered and treated independently of the hydronephrosis, which will resolve when the obstruction has been relieved. *Bilateral types*

A small number of bilateral cases are seen in which the pathological cause is identical with that of the unilateral type, as in the instances of bilateral renal calculi, or of the bilateral dynamic dysfunction already mentioned. The remaining unilateral cases arise from a variety of causes: those within the lumen of the renal pelvis and ureter, for example, stone or papilloma; those in the wall of the ureter, such as stricture or carcinoma; and extrinsic pressure effects from such conditions as malignant growths, diverticulum of the bladder and abnormal renal vessels. *Unilateral types*

3. SURGICAL ANATOMY

An unusual arrangement of the blood-vessels of the renal pedicle is sometimes associated with a hydronephrosis, but in the majority of cases this is a coincidence rather than a primary cause (Fig. 123). *Renal pedicle*

4. PATHOLOGY

The changing contour of the renal pelvis and calyces during their early enlargement follows one of two lines: in one the major and minor calyces are primarily affected, whereas in the other the dilatation is of the renal pelvis itself. *Contour of pelvis*

In renal calculus, a common cause of the obstructive type of hydronephrosis, the renal pelvis is frequently within average limits whereas the calyces show gross dilatation, and only in the later stages does this dilatation affect the pelvis proper.

This feature may be contrasted with the so-called dynamic case in which the renal pelvis may be markedly enlarged without dilatation of the calyces (Fig. 124); it is true that at a later stage the latter do increase in size, but secondary mechanical obstructive factors have here become superadded, and with them their natural sequelae. *Dynamic factor* *Superadded mechanical factors*

Such divergent features in the early phase of the two groups suggest different underlying causes. Those of the first are characteristic of organic obstruction, whereas in the second group the early changes are more closely comparable with those seen in achalasia of the cardia and Hirschsprung's disease. If a ureteric catheter passing into the renal pelvis is connected to a water manometer and recording drum, it can be shown that the pressure inside one

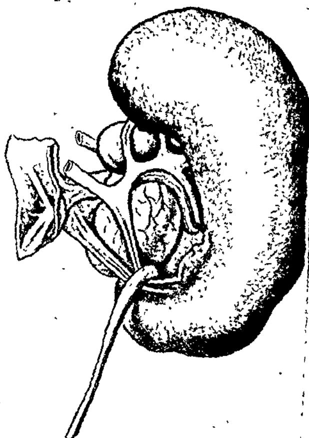


FIG 123.—The anatomical relation between the renal pedicle and pelvi-ureteric region, showing the ureter passing over an abnormal vessel.

of the dynamic hydronephroses is quite low while the pelvic muscle is inert. When sympathetic impulses to the kidney are blocked by a spinal anaesthetic up to the level of the fifth thoracic segment, the previously inert pelvis now shows rhythmic contractions (Fig. 125), while the intrapelvic pressure rises. Apart from the light which it throws on the nature of the hydronephrosis, this test is valuable in deciding when a case is suitable for treatment by renal sympathectomy (Underwood, 1937) (see p. 290).

The part played by abnormal renal vessels has been well described by Winsbury-White (1925) who considers that the enlarging renal pelvis bulges forwards and downwards, taking with it the upper portion of the ureter. This enlargement of the renal pelvis similarly accounts for the high pelvi-ureteric junction which produces a valvular effect. This mechanism will clearly be accentuated when the ureter is pressed against the renal pelvis by an abnormally placed vessel.

When this anatomical arrangement is present without an abnormal vessel, examination of this area often shows the sequelae of past inflammatory changes in the form of adhesions in the pelvi-ureteric space. The local stretching injury at this region, caused by the attacks of colic in these cases, will account for the fibrosis.



FIG. 124.—Showing the pyelogram of a case suitable for pyeloplasty; note that the minor calyces are not markedly dilated, indicating little diminution of renal cortex in spite of the large hydronephrosis

Stricture

Round-celled infiltration at the site of a physiological sphincter is well seen at the pelvi-ureteric junction in

dynamic hydronephroses, and ultimately recurrent inflammation causes true organic obstruction—a fibrous stricture.

5. CLINICAL PICTURE

The essential features are those of renal colic with tenderness on bimanual palpation of the kidney. In more advanced cases enlargement of the renal pelvis may be palpable. Haematuria is not uncommon and pyrexia indicates a superadded infection of the urine.

When the hydronephrosis is primarily of the mechanical obstructive type, there will be coincident clinical features of the primary cause, for example a stone, or growths of the ureter or bladder.

In the dynamic cases, however, and especially in the early stage when no secondary mechanical factors have become superadded, the salient picture is of attacks of renal colic. These are cases in which early diagnosis, before the renal cortex is damaged, provides the opportunity to treat the condition while it is still curable.

6. SPECIAL AIDS TO DIAGNOSIS

Radiography

The taking of a skiagram is the first line of investigation, and thus a calculus may be diagnosed as the cause of renal colic with or without hydronephrosis.

(1) Intravenous pyelography

In addition to demonstrating the outline of the renal pelvis and calyces, intravenous pyelography gives valuable information regarding kidney function. A dense shadow need not necessarily indicate good kidney function, for the shadow of a kidney pelvis having a stone impacted at its outlet may be good, although the renal cortex is in process of destruction. Similarly, in a large dynamic hydronephrosis having good renal tissue, the excreted dye may be so diluted as to cast little or no shadow. In this case, however, it is not uncommon for the intravenous pyelograph to show rounded shadows—the cups of the minor calyces in which the dye is lying.

(2) Instrumental pyelography

Dangers

Instrumental pyelography may be used in cases in which adequate evidence is not available by the intravenous method, but it should be undertaken only with care and by an experienced operator.

Difficulties

Occasionally, the ureteric catheter may not pass into the pelvis; the sodium iodide solution may then flow back down the ureter or a small quantity may pass into the pelvis, which is very suggestive of a hydronephrosis with the ureter leaving the pelvis parallel to its wall.

Precautions

Over-distension of the pelvis with sodium iodide solution should be rigidly avoided since it may be followed by untoward sequelae. This can be avoided by serial x-ray photographs, the first being viewed after a small quantity of solution has been injected; as a further precaution a large quantity of potentially irritating solution should not be left in the renal pelvis free to permeate through the collecting tubules into the renal cortex—it should be drawn off before withdrawal of the ureteric catheter.

(3) Pressure tests

Effects of spinal anaesthesia

Pressure tracings are of value in determining whether a case is suitable for renal sympathectomy, for this treatment is unsuitable unless it can be shown

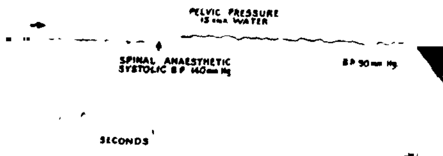


FIG. 125.—Tracing showing that the inert renal pelvis contracts rhythmically following a spinal anaesthetic.

that the renal pelvis still retains its power of contraction, as demonstrated by spinal anaesthesia (Fig. 125).

7. DIFFERENTIAL DIAGNOSIS

There are conditions which may give rise to a swelling having the physical characters of hydronephrosis but which are not associated with attacks of renal pain, whereas the commonest cause of renal colic is a stone, which is usually demonstrable by x-ray examination.

In brief, therefore, the diagnosis of hydronephrosis should always be considered when there is renal colic without a stone, when the kidney is not abnormally mobile, and whether the enlarged renal pelvis is palpable or not.

8. PROGNOSIS

Renal damage

Bilateral hydronephrosis

In bilateral cases due to abnormalities of the lower urinary tract hydronephrosis indicates the severity of the primary condition and the outlook is therefore serious. The prognosis depends not so much upon the kidney as upon the general health of the patient. Usually the outlook is not good, but if adequate treatment is possible it is surprising how such cases do improve.

Infection

Infection, especially when recurrent, is an important feature, but this commonly improves *part passu* with the hydronephrosis.

Unilateral hydronephrosis

In unilateral cases due to acquired causes the prognosis depends upon the response of the causative condition to treatment; provided that there has not been undue damage to the kidney cortex, the hydronephrosis diminishes and the prognosis is good.

Favourable cases

In the dynamic form the most favourable cases are those in which (1) the major calyces are not markedly enlarged, (2) the renal pelvis still retains its contractility, and (3) there is no superadded mechanical obstruction due to one of the complications of this type. Such complications are, for example, pressure by abnormal renal vessels, back-pressure by lateral implantation of the ureter giving a valve effect, and thickening at the pelvi-ureteral junction.

Groups (1) and (2) can be accurately assessed by special aids to diagnosis; group (3) may be suspected but can be accurately assessed only at operation.

9. INDICATIONS FOR SURGERY

These may be classified under two headings:

(1) Treatment of the cause.

(2) Treatment of the hydronephrosis. There are certain primary aims: these are (a) reduction in size of the enlarged pelvis; (b) to ensure adequate drainage of urine from the renal pelvis.

Secondary aims are curtailment of the progressive destruction of the renal cortex and prevention of urinary infection.

Treatment of extrinsic causes

Generally speaking the condition is progressive, and surgery offers the patient the best chance of recovery. In the small group in which operation upon the kidney is not indicated—the bilateral type secondary to obstruction of the lower urinary tract—the treatment is that of the primary condition.

10. PRE-OPERATIVE MANAGEMENT OF THE PATIENT

Sterility of urine

It is important that the urine should be sterile before operation, especially when a plastic procedure is contemplated. In most cases infection will respond to a course of chemotherapy and free diuresis, but occasionally a preliminary nephrostomy is necessary.

Renal function

When the condition is advanced and especially if it is bilateral, the function of both kidneys and the state of the blood chemistry should be assessed beforehand, since the advisability of removing one kidney may have to be contemplated.

11. OPERATIVE TECHNIQUE

Having regard to the aims of operation already described, it can be accepted that ligation of aberrant vessels alone is inadequate and that removal of a wedge of renal pelvis will do no permanent good.

Depending upon operative findings, the following surgical procedures are used either singly or together.

(1) Renal sympathectomy

Su[illegible] cas[illegible] *Ca[illegible] fail[illegible]*

In dynamic cases in which no superadded mechanical obstructive factors are present, or in which such factors can be adequately treated at the time, this operation gives good results if it is performed early, before the muscle of the renal pelvis has atrophied or become fibrosed. At operation any superadded secondary mechanical factors are usually evident, but special attention should be paid to thickening at the pelvi-ureteric junction, for sympathectomy cannot be expected to succeed in the presence of such complications.

Ligature of aberrant vessels

The mechanical factor may, however, be minimal and then, in addition to renal sympathectomy, it may be sufficient to free adhesions at the pelvi-ureteric junction or to divide aberrant renal vessels. The latter procedure, especially when the vessels are large, should be undertaken with caution in view of the danger of subsequent infarction of the lower pole.

On theoretical grounds the best end-result would follow blocking of sympathetic impulses passing to the renal pelvis and upper ureter, leaving intact

parasympathetic fibres coming, in all probability, down the vagus. Th could be achieved by resection of the splanchnic nerves, but this is a maj procedure and there is as yet insufficient evidence to prove that the end-resi is better than that of denervating the renal pedicle.

End-result

After renal sympathectomy it has been shown that the pelvis empt rhythmically under its own intrinsic neuro-muscular mechanism while the large sac of stagnant urine is changed to a smaller cavity approaching that of a normal renal pelvis.

The technique of this operation is described elsewhere. (*See* Kidney and Ureter—Denervation of Kidney, p. 159.)

(2) Pyeloplasty

Th ... operation upon the renal pelvis are: ... mechani- ... e ... 1.

(1) ... pel...

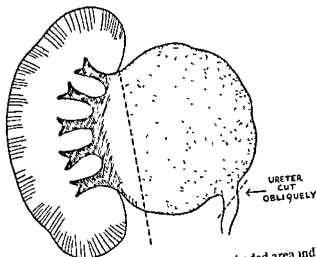


Fig. 126 —Stage 1 of pyeloplasty, the shaded area indicates the portion to be removed

The fundamental aims of the operation are reduction in size of the pelvis, more rapid emptying of urine into the ureter, and therefore diminution of residual urine in the renal pelvis.

Excision of pelvis and ureter

(*a*) *Stage 1: removal of redundant pelvis*

Enough pelvis is excised to reduce the cavity t an average size, with a anatomical contour as ne to normal as possible. Ca is taken to ensure that t.. lowest part of the opening is at the lowest part of the pelvis; this ensures complete drainage (Fig. 126).

Nephrostomy and splint catheter

(*b*) *Stage 2· temporary nephrostomy and insertion of splint catheter*

For the nephrostomy a Malecot catheter, size 14, Charière is recommended; for the splint a ureteric catheter will be found satisfactory, for it is of adequate calibre but not so large as to cause pressure at the anastomosis line, which encourages subsequent stenosis.

Technique of suturing

(*c*) *Stage 3: anastomosis of ureter to dependent part of renal pelvis*

Interrupted mattress sutures of No. 000 plain catgut are used with the loop on the muscle coat, thus avoiding the mucosal ledges, which are liable to cause obstruction when the ureter is implanted direct through a stab wound into the renal pelvis (Fig. 127).

Reconstruction of renal pelvis

(*d*) *Stage 4: closure of blind end of renal pelvis*

This may be done with a continuous suture (Fig. 128). At the pelvi-ureteral anastomosis it is wise to insert a few additional stitches to take the strain

off the internal layers of sutures, the function of which is essentially that of accurate apposition.

(e) *Stage 5· nephropexy*

This fixes the kidney, which will now be found to have an unusually long pedicle. *Nephropexy*

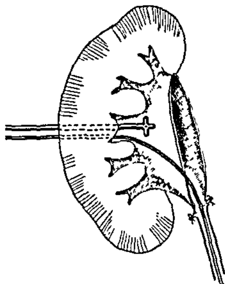


FIG. 127.—Stage 2 and stage 3 combined.

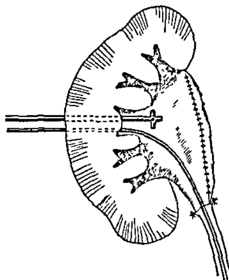


FIG. 128.—Stage 4 completed.

The lumbar wound is closed, leaving a light corrugated rubber drain down to the site of the anastomosis.

(3) Nephrectomy

Removal of a kidney for hydronephrosis is justified when there is so little renal cortex remaining that, in the presence of a sound kidney on the other side, the function of the diseased kidney is negligible.

12. POST-OPERATIVE CARE

Cases of renal sympathectomy and nephrectomy call for no special urological after-care.

The after-care of pyeloplasty is important, since premature passage of urine down the ureter may cause troublesome leakage. Although each case should be treated on its special merits, the following may be taken as a guide.

(i) *Splint catheter.*—Irrigate daily with a few cubic centimetres of saline solution to keep the ureter clear of debris, and remove the catheter on the fifth day.

(ii) *Wound drainage.*—The corrugated rubber drain is shortened on the second day, and removed on the third day *Wound drainage*

(iii) *Nephrostomy.*—Upon free and unobstructed drainage of the nephrostomy tube depends the whole success of the operation. It should be kept clear by irrigation with saline solution twice daily, using not more than ½ ounce of saline solution at each injection. It must be shown that the fluid runs back freely before a further quantity is run in. On or about the tenth day patency *Test of reconstructed channel*

of the reconstructed channel may be proved by running in a little methylene-blue solution and occluding the tube, when the colour should appear in the next specimen of urine passed. The tube may then be removed, and the nephrostomy wound will usually dry up in 48 hours.

13. RESULTS OF TREATMENT

After-progress of cases can readily be followed by intravenous pyelography.

Post-operative stricture

Stricture formation at the pelvi-ureteric junction following the plastic procedure is a complication which causes infection, and which may necessitate nephrectomy. It is avoided by careful technique and selection of cases.

Conservative surgery as opposed to nephrectomy is justified in the treatment of hydronephrosis; it may be successful but, if it is not, nephrectomy is still available as a secondary instead of as a primary operation.

PART II
PYONEPHROSIS

1 DEFINITION

A pyonephrosis is a dilated kidney filled with pus, commonly the organ is also enlarged to form a palpable swelling. There are all degrees between a suppurative pyelonephritis and an infected hydronephrosis, though strictly speaking *the term, pyonephrosis, is applied only to a kidney in* which the cortex has been excavated, the resulting cavity being filled with pus.

2. AETIOLOGY

Common causes

There are three common causes—calculus, tuberculosis and infected hydronephrosis. Tuberculosis of the kidney is described on pp. 307 to 324. The remaining two conditions are further subdivided into the so-called open or closed types; in the former, pus escapes and drains down the ureter, whereas in the latter it is temporarily held up in the kidney.

3. PATHOLOGY

In the early stage the calyces become distended, the pyramids disappear and the mucous membrane is ulcerated; later, abscesses form in the renal cortex;

these communicate with the renal pelvis, which is now lined with granulation tissue and is filled with pus and debris, including phosphatic deposits.

Perinephric spread Inflammation of the perinephric tissues causes dense fibrous adhesion to all surrounding structures, notably to viscera, blood-vessels and muscles. In late cases the suppuration may spread into surrounding tissues to form a perinephric abscess.

Organisms present The organisms present are varied. *Bacillus coli* is an early arrival, and later staphylococci appear, while at times anaerobic organisms may be found.

4. CLINICAL PICTURE

The symptoms and signs are usually constant pain in the loin, with pyuria, and an enlarged kidney. The whole clinical course is prone to exacerbations associated with increase in the lumbar pain, pyrexia and tenderness in the loin.

Exacerbation of symptoms Frequently these exacerbations are ushered in by a diminution of the pyuria and an increase in the size of the swelling. In effect the onset of one of these exacerbations often indicates a change from the open to the closed type of pyonephrosis.

5. SPECIAL AIDS TO DIAGNOSIS

Cystoscopy Although the clinical picture is usually definite, examination of the urine, radiography, intravenous pyelography and cystoscopy give valuable ancillary evidence. The pyuria is usually macroscopic and at cystoscopy pus may often be seen exuding from the affected ureter.

Radiography and intravenous pyelography A skiagram will show any calculi present. Intravenous pyelography gives evidence of gross destruction of the affected kidney with diminution or absence of function, and will also provide useful information about the function of the opposite kidney.

6. DIFFERENTIAL DIAGNOSIS

Pyuria Perinephric abscess and carbuncle of the kidney give similar clinical features; in both conditions, however, pyuria as a rule is absent and, if present, is seen only in microscopic amounts.

Oedema of abdominal wall Deep oedema is often found in the lumbar abdominal wall if a perinephric abscess is present, whereas in uncomplicated cases of carbuncle and pyonephrosis this is not seen.

Outline of swelling The general outline of the swelling is palpable in a case of pyonephrosis or of carbuncle of the kidney, whereas in perinephric abscess this is more difficult to determine owing to the diffuse oedema of the surrounding tissues.

7. PROGNOSIS

Unilateral and bilateral types When the condition is bilateral the outlook is not good, but if the disease is confined to one kidney the patient has every chance of complete recovery, provided that nephrectomy is possible and that the function of the remaining kidney is adequate.

Indications for surgical intervention

Resolution rare It is so rare for an established pyonephrosis to resolve or even to remain static that radical measures are recommended.

8. PRE-OPERATIVE MANAGEMENT

Urinary function

Anaemia

A routine assessment of urinary function is important and, in view of the *anaemia which is frequently present, a blood transfusion prior to operation* is of value.

Preliminary nephrostomy

In a small number of cases the general condition is so poor that a preliminary nephrostomy is all that the patient will stand; this, however, should be *done only when really necessary, for it* introduces the dangers of possible secondary haemorrhage and adds difficulties to the subsequent nephrectomy.

9. OPERATIVE TECHNIQUE

Adequate exposure

Nephrectomy should be carried out through an incision of sufficient length to give generous exposure, for the organ is enlarged, adherent and often friable, so that its removal usually presents great difficulty.

Intracapsular nephrectomy

If adhesions to the peritoneum and to other important structures are unusually dense it is safer to carry out an intracapsular nephrectomy which, though it may involve bleeding from the friable kidney obviates the dangers of damage to viscera and blood-vessels, particularly to the inferior vena cava.

Ligature of renal pedicle

The renal pedicle is usually an oedematous mass of fibrous tissue, and the vessels should be secured and ligated with care.

Not infrequently the renal cortex and pedicle are so fibrosed that the vessels are small, and then if haemorrhage occurs it is not a major problem.

The cavity is dusted with sulphanilamide powder and the wound is closed with drainage.

Conservative measures

Rarely conservative measures can improve the pyonephrosis, as in the case of a single removable calculus, but conservatism is indicated only when the condition of the opposite kidney prejudices the result of nephrectomy.

Permanent nephrostomy

When the condition is bilateral, nephrostomy may be needed as a life-saving measure

10. POST-OPERATIVE CARE

Chemotherapy

A course of chemotherapy is valuable during the early post-operative days until the temperature has settled and the operation area appears clean; there is frequently a sero-purulent discharge for several days, but this is of no serious import.

11. RESULTS OF TREATMENT

[illegible] improve [illegible]

and the remaining kidney is sound, the patient regains good health [illegible] short time.

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KIDNEY AND URETER—STONE

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1. DEFINITION

212.] Kidney stones are formed by crystalloids, normally held in solution by a colloid, being precipitated and then bound together by a cement-like substance. Errors in metabolism—congenital, as in cystinuria, or acquired, as in uric aciduria—may produce crystalluria; stagnation will both predispose to crystal formation and allow these crystals to be bound together into a stone.

Renal calculi are more common in men, because in women, particularly during menstruation, there is a higher rate of excretion of citric acid which is a solvent (Robinson, 1947a).

Diet, by high ingestion of oxalates; climate, by diminishing fluid output, hypervitaminosis D and hypovitaminosis A are factors in stone production. Infection and stagnation, as in hydronephrosis, and particularly in patients kept in a recumbent position, are powerful predisposing causes.

Types of stone

(a) Phosphate stones—amorphous and almost invariably associated with staphylococcal infection which produces alkaline urine by splitting urea into ammonia—are radio-opaque.

(b) Triple phosphate stones, which are usually sterile, are recognized radiologically as jackstones, and by coffin-lid crystals in the urine.

(c) Calcium oxalate stones, occurring usually in sterile acid urine and showing envelopes and dumb-bell crystals in the urine, are radio-opaque.

(d) Uric acid stones in older gouty patients show acicular crystals in a very acid urine and are not radio-opaque.

(e) Stones containing cystine, xanthine and other substances occur rarely.

Secondary infection

All these stones may be found with secondary urinary infection and deposits of phosphates and urates. *Bacillus proteus* is often associated, but almost any bacteria may be present. Stones may occur in association with other renal diseases, as in carcinoma of the renal pelvis and in tuberculosis. Ureteric stones are usually migrated kidney stones, and only rarely form in the ureter, for example, behind a stricture—as in ureterocele. They are aetiologically similar to kidney stones.

2. SURGICAL ANATOMY

Stones usually form in the dependent lower calyces, or in the upper calyces of recumbent patients, growth taking place into the pelvis and other calyces. Migration occurs into the pelvis. The stone may be held up there or in the ureter, particularly above the narrow uretero-pelvic and uretero-vesical area.

3. PATHOLOGY

The stone may produce local hydronephrosis of one calyx, of the pelvis and calyces, and of the ureter, according to the site of the obstruction; the cortex is thinned and, in a long-standing case, fat replacement is marked in the medulla and is associated with fibrosis and thickening of the pelvis itself.

Pyelonephritis and abscess formation

Infection hastens this process, leading to pyelonephritis and abscess formation in the kidney and perinephric tissues. Most cases give evidence of past perinephritis by the presence of capsular adhesions. These may involve the diaphragm, the muscles of the posterior abdominal wall, and the colon, and on the right side additionally the duodenum and the vena cava. Liver and spleen are not adherent to the kidney in stone cases. There may be some ulceration at the site of the stone impaction, but subsequent healing of the scar tissue rarely leads to ureteric stricture.

Some degree of reflex atonia in the contralateral renal pelvis and ureter is usually observed. Vascular changes in the renal vessels may lead to hypertension, even with unilateral disease.

4. CLINICAL PICTURE

Pain

Fixed pain varies with the level of the stone. When it is in the renal substance, pain is felt posteriorly in the renal angle; if in the pelvis, anteriorly at the same level; and during the passage down the ureter, pain is felt in the iliac fossa and is referred segmentally to the inner side of the thigh and genitalia. In the last part of the ureter it produces a sensation as of bladder stone, with pain at the end of micturition.

Colic is associated with spasms at the time of the stone migration and agonizing pain with vomiting, which is often not relieved by morphine and is accompanied by guarding of the abdominal muscles.

Haematuria

Bleeding may be severe, bright red and symptomless. With large-branched calculi, it may simulate bleeding from a neoplasm, with the formation of ureteric blood casts. More commonly, short attacks of bleeding coincide with stone migration and may be associated with exercise.

Anuria may result from bilateral calculus obstruction or from obstruction of a solitary kidney. More rarely it results from the obstruction of one kidney and reflex suppression of secretion on the other side. *Anuria*

Evidence of incipient uraemia—headache, dyspepsia, diarrhoea and secondary anaemia—may be found in cases of bilateral calculi; oedema and fits are not common. *Uraemia*

Associated symptoms of pyonephritis may occur at any time, with a temperature of 104° F., severe backache and acute uraemia, and will give rise to marked tenderness over an enlarged kidney. The tongue is furred. The level of abdominal tenderness gives some guide to the position of the stone, which may sometimes be felt, per rectum, above the prostate gland or, *per vaginam* at one side of the cervix. *Infection*

Pyuria will vary but the presence of stone and obstruction will give rise to pyonephrosis from which drainage will be "closed".

Increased frequency of micturition may be reflex; it may also result from associated urinary infection or from irritation by the passage of crystals. *Frequency of micturition*

5. METHODS OF INVESTIGATION

(1) Radiography

Intravenous pyelography is of first importance, but emphasis must be given to the need for a plain skiagram before injection, so that a stone is not masked by the "dye"; skiagrams at 5 and 15 minutes after injection suffice in most cases, but additional skiagrams after 30 and 60 minutes may be necessary in cases in which kidney function is impaired. *Intravenous pyelography*

A shadow of a stone will be seen in the preliminary skiagram; its relation to calyces, pelvis and ureter and a good idea of the function of the contralateral kidney is obtained from the later skiagrams. The function of the affected side must be considered in relation to the length of the history, since, in a previously healthy kidney, complete obstruction of up to 6 weeks' duration may later be restored to normal, giving a normal excretion pyelogram. A uric acid stone may appear as a filling defect and may adsorb the "dye" in later pictures.

Lateral pictures are often useful in differentiating between calculi and calcified glands in the renal area in doubtful cases, stones lying on a level with the bodies of the vertebrae and the gland lying well in front. It must be remembered, however, that, in a large hydronephrosis, a stone may be lodged in a calyx far anteriorly in the mass. *Calcified glands*

(2) Cystoscopy

Cystoscopy is next indicated so that a specimen of urine may be obtained for examination for organisms, and in order to confirm the diagnosis. If the stone has been recently voided, the orifice of the ureter concerned may show evidence of the trauma by small submucous haemorrhages in the immediate neighbourhood A stone lodged near the orifice may produce oedema around the opening and may be seen actually peeping out, and any predisposing cause such as ureterocele will be noted and, on occasion, treated. In advanced pyonephrosis, a rigid dilated ureteric orifice may need careful investigation to exclude tuberculosis. *Ureterocele*

The efflux should be observed; it may be blood-stained, turbid or absent.

(3) Retrograde pyelography

Retrograde pyelography will be carried out at the time of the cystoscopy when sufficient detail is not provided by the excretion urogram, mainly when there is doubt about the shadow being a ureteric stone, in cases in which the ureter does not fill well with "dye" and when complete obstruction or kidney damage causes inadequate secretion. The level of any obstruction to the *opaque ureteric catheter is noted and* a preliminary picture is taken to show the relationship to the catheter, as well as to ensure that the catheter is not too high in the kidney, thus requiring a slight withdrawal before the injection of the 13 5 per cent solution of sodium iodide. A bilateral retrograde pyelogram should be carried out simultaneously only in exceptional cases.

(4) Estimation of renal function

Bilateral impairment of function

On the whole, sufficient information is given by the intravenous pyelogram, but when function of both sides is impaired, useful comparison may be obtained by intravenous indigo-carmine (10 cubic centimetres of 0 4 per cent solution), secreted normally in 3 minutes, light blue, becoming dark blue in 6 minutes.

Blood-urea estimation is useful in assessing progress in uraemic patients so *that the most favourable time may be chosen for operation. Further information* may be gained by the urea clearance and urea concentration tests.

6. DIFFERENTIAL DIAGNOSIS

(1) Appendicitis and appendicular colic present the greatest clinical difficulty, and many an appendicectomy is performed for ureteric stone. In the doubtful cases radiography is the best safeguard.

(2) Abdominal adenitis, including calcified glands, may produce very similar pain; this must be recognized and excluded by pyelography as well as by the greater mobility of the more mottled shadows with clear margins.

Tubal gestation

[illegible]

pain, however, may be indistinguishable from ureteric colic.

Tuberculosis

(4) Renal calcification can be produced in tuberculosis, which should be recognized by the presence of bacilluria and by the radiological and cystoscopic appearances. *Renal neoplasm, often calcified diffusely throughout,* will show deformity of the kidney outline or of the calyces. Kidney cysts calcify at the periphery, giving a circular outline.

(5) Hyperparathyroidism will produce intratubular calcification, appearing bilaterally far in the renal substance; these are rarely stones but are of special aetiology.

Gall-stones

(6) Gall-stones are far anterior to the kidney in a lateral picture but, for complete localization, these may require cholecystography as well as pyelography.

(7) Phleboliths are symptomless round shadows in the neighbourhood of the lower ureter; they have a smooth margin and are nearly circular, the larger ones having a less opaque centre; they tend to *occur in lines curving* to the ischial spines. Passage of an opaque catheter may be necessary to prove the shadow to be extra-ureteric.

7. MEDICAL TREATMENT

Colic is best relieved by papaverine, 2 grains, and by injections of Trasentin, 1 cubic centimetre twice daily, to relax the ureter and to assist the natural passage of a stone. In a surprisingly large proportion of cases, stones are passed naturally and the prognosis as to this chance may be given by the relative size of the stone and of the ureter in the excretion urogram

Natural passage

Natural passage should be encouraged, between bouts of colic, by exercise, such as skipping, by administration of antispasmodics (Trasentin) and by diuresis. Recumbent patients should have their positions altered (Pugh, 1933). Most spinal and hip cases can be nursed in the prone position for a week or two (Pulvertaft, 1939). The descent and progress of the stone must be assessed by intravenous pyelography at intervals of 14 days, and all urine should be voided into an easily examined vessel so that the stone is not overlooked.

Associated infection must be treated by chemotherapy and penicillin, but in association with urinary obstruction it is an indication for urgent operation, because such a kidney may be very rapidly destroyed

With diuresis and diet, bilateral cases unsuitable for operation may continue for many years without any increase in the size of the stones. This also applies to symptomless stones smaller than a pea, which may be difficult to find at operation.

Solution G1 (Citric Acid Monohyd. 32·25 grammes, Magnes. Oxid. Anhyd. 3·84 grammes, Sod. Carb. Anhyd. 4·37 grammes, Aq. Dist to 1,000 cubic centimetres) has not fulfilled its promise of dissolving renal stones by lavage through an indwelling ureteric catheter (Suby, 1944).

8. SURGICAL TREATMENT

(1) Pyelolithotomy

When the kidney is worthy of preservation, by virtue of good function, absence of pyonephrosis, no previous removal of stone, or owing to disease of the opposite kidney, pyelolithotomy offers the best approach for the majority of cases.

Indications for operation

Approach to the kidney and upper part of the ureter.—The patient lies on his side with the lower leg flexed at the hip and knee, with the top leg straight, and a sand-bag placed between the bottom ankle and the top knee. The kidney bridge is raised or a sand-bag is placed between the ribs and iliac crest to open the loin as wide as possible. The patient can be kept in position by straps

The curved lumbar incision from the angle between the erector spinae and the twelfth rib to 1½ inches above the anterior superior spine is most generally favoured; this may be prolonged anteriorly if necessary. The skin is slightly undercut in view of subsequent retraction of divided muscles, and the external and internal *oblique muscles are cut across, the transversus being split in the* direction of its fibres after pushing forward the peritoneum in the anterior [illegible]

dislocated upwards, care being taken not to injure the pleura which should be held off by a finger at the time. The perinephric fat should be incised low down to avoid the pleura, and well posteriorly to avoid the peritoneum, and the kidney then cleared as may be necessary.

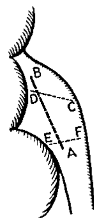


FIG. 129.—Incisions in the renal pelvis Incision A–B affords the best approach, with C–D there is a risk of bleeding if it is prolonged at D; with E–F there is a risk of avulsion of the ureter

The pelvis is exposed and two sling guides are inserted to facilitate the incision in the pelvis; this is best made in the longest axis but must not be prolonged on to the kidney since troublesome haemorrhage occurs from vessels at the edge of the renal substance. If made transversely at the edge of the renal substance across the pelvis, care must be taken not to extend the incision by tearing during removal of the stone, because the ureter may be avulsed (Fig. 129).

Numerous stone forceps have been designed but Thompson-Walker's are most generally useful. After removal of the stone any debris may be washed out by irrigation with silver nitrate 1 : 10,000, which is also useful to diminish bleeding. A ureteric catheter must be passed to prove the patency of the ureter. One or two sutures are usually sufficient to close the incision in the pelvis, which may be left unsutured (Robinson, 1947b) but covered with fat in difficult cases. The wound must be drained for 1 week to allow the escape of urine.

(2) Nephrectomy

Nephrectomy is the operation of choice for stones associated with gross kidney damage, pyonephrosis or previous stone formation, provided that the other kidney is healthy and likely to remain so. In recumbent cases operation should be deferred until the patient is ambulatory, even if this means a temporary nephrostomy in the event of pyonephrosis

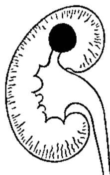


FIG. 130.—A stone in renal substance with a narrow communicating channel. The stone is best removed by nephrolithotomy, since there is a risk of trauma and bleeding if it is removed through the pelvis.

(3) Nephrolithotomy

Nephrolithotomy may cause more damage to the kidney, and is accompanied by more bleeding than is the case with pyelolithotomy; there is also some tendency to haemorrhage on the fourth day on absorption of the sutures. It is indicated for stones isolated in a calyx with a narrow communicating channel to the pelvis, and in cases in which the pelvis is absent or largely intrarenal (Fig. 130). In branched calculi, nephrolithotomy may need to be combined with pyelolithotomy, and a finger in the renal pelvis may be a helpful guide to the cortical incision.

After locating the stone by palpation or, if necessary, by probing through the outer border of the kidney with a fine straight needle, the convex border of the kidney is incised down to the stone, bleeding being temporarily

controlled by digital pressure on the pedicle, by the other hand or that of an assistant. Bleeding is controlled by mattress sutures introduced through deep bites of the renal substance.

Closure of wound

To close the wound after any of these operations upon the kidney, interrupted sutures should be introduced through the muscles and tied after removal of the kidney bridge or sand-bag.

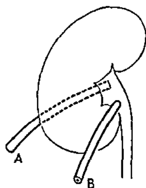


FIG. 131.—Nephrostomy tube (A) at the lower pole of the kidney, a second tube (B) may also traverse the kidney substance or pass, as shown, directly to the renal pelvis.

(4) Nephrostomy

The insertion of a tube (No. 20 French) through the kidney substance into the pelvis for a few days is said to minimize the risk of later haemorrhage. Nephrostomy is also of value in cases of infection and again with a patient recumbent as a result of other diseases (for example, tuberculous hip) in whom an operation for stone cannot be delayed until he is ambulatory. To prevent stone re-formation the tube will need to be retained over a period of 6 months. The indications for intervention in recumbency cases depend on three main factors—the rate of growth of the stone, the degree of associated renal damage from obstruction, and the supervention of infection; when renal deterioration is not occurring operation is preferably delayed until the patient is no longer recumbent. The tubes should be connected to drainage bottles at the bedside.

Bilateral calculi

The irrigation of a kidney with Solution G1 (Keyser, 1943) will dissolve phosphate stones but it has been found to be ineffectual when introduced through a ureteric catheter, owing to the small surface irrigated and the length of time required —usually many weeks. Nevertheless irrigation through a nephrostomy tube—or better still, through two nephrostomy tubes, one for inlet and one for outlet—is very efficient in dissolving relics which might be missed in the removal of a branched calculus (Fig. 131). In the past, any fragments left behind would have led to rapid stone re-formation, but with this safeguard it is now possible to operate on bilateral calculi which were previously considered to be unsuitable for surgery. The irrigation is given by constant drip for some weeks until there is no radiological evidence of debris, and is only discontinued if, after stopping for an occasional 24 hours, there is no recurrence of pain. The irrigation in the first kidney should be continued until the second kidney has also been effectively cleared of debris and infection by similar means.

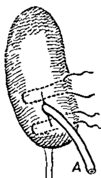


FIG. 132.—Mattress sutures placed widely to control bleeding after insertion of nephrostomy tube (A)

Pyonephrosis

Nephrostomy may be needed as a prelude to nephrectomy in calculus pyonephrosis to drain the pus and to allow the patient's general condition to

improve. Nowadays, with chemotherapy and penicillin available, this is needed less frequently.

Nephrostomy tubes are best placed at the lower end of the kidney to avoid kinking (Cabot and Holland, 1931), and should be stitched in at first (Fig. 132). If the tube slips out it should be replaced at once, since the sinus contracts very rapidly.

Bilateral nephrostomy may be needed for calculous anuria, and is best carried out through two parallel (Edebohl) incisions, with the patient lying on his face (Fig. 133).

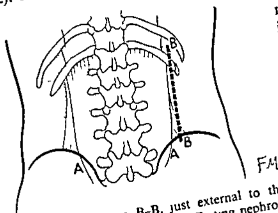


FIG 133.—Incision B–B, just external to the sacrospinalis, is useful for effecting nephrostomy as both sides may be done through two incisions without altering the position of the patient on the table. A–A lateral border of sacrospinalis on each side.

(5) Ureteric stones: endoscopic methods

To assist the natural passage of ureteric stones numerous instruments have been devised and success has been claimed for each, but they are not entirely safe (Dourmashkin, 1945). Two procedures are, however, well worth a trial.

(i) *Ureteric catheter.*—Ureteric catheterization up to and beyond the stone, with one or more ureteric catheters, may dilate the ureter below or shift the stone so that it is passed more readily. This procedure is usually accompanied by the injection of liquid paraffin, 1 cubic centimetre, as a lubricant.

(ii) *Meatotomy.*—When the stone is impacted above a tight meatus, incision of the orifice for about 1 centimetre through a cystoscope is of very great benefit. This is best done by a meatotome, using the endothermy current, but a stilette may be threaded through a ureteric catheter to protrude 0 5 centimetre, and the proximal end connected, with careful insulation, to the diathermy lead, the catheter and stilette being passed down the cystoscope as a meatotome. Bleeding will occur if the incision is of more than 1 centimetre.

Ureterolithotomy

Open removal of the stone from the ureter is indicated by unlikelihood, o failure, of any descent, by failure of endoscopic methods, or by superventio of infection or of renal failure.

Incision

On the way to the theatre, the patient should have a further x-ray examination to detect any change in the position of the stone. Above the sacro-iliac joint the ureter is exposed through the curved kidney incision, and located lying on the posterior wall behind the peritoneum. To avoid losing the stone, a tape sling should be passed around the ureter above the stone. A small incision is made in the long axis of the ureter above the impacted stone if it can be milked up, but otherwise it is made over the stone, which is peeled out of the mucosa.

Aspiration of infected urine

Any infected urine escaping is aspirated by a sucker, but attempts to prevent escape by digital occlusion from above are worth while. The patency of the lumen below is proved by passing a ureteric catheter down

the ureter. The ureteric incision is closed by one or two Lembert's sutures and the wound is drained for 1 week.

Stones below the sacro-iliac joint are best approached through a median sub-umbilical incision separating the recti and carefully pushing the peritoneum upwards and inwards off the lateral pelvic wall. The ureter is located as it dips into the pelvis and then crosses the iliac vessels, but it may be pushed inwards with the peritoneum. A tape sling should be used as above, and if the stones cannot be milked up to an accessible region, it may be necessary to divide between clamps some vesical vessels crossing the ureter, in order to gain approach to the stone. It is advisable to have the bladder empty. Incision and removal of the stone and drainage are as for the lumbar ureterolithotomy. Ureteric stones should not be approached through the bladder however tempting this method may seem.

(6) Calculous anuria

Calculous anuria as a primary symptom is very rare, but may occur more commonly in patients with stone even when under treatment, as for example when a recumbent patient becomes ambulatory and much grit and calculous material are voided, with resultant obstruction to both ureters in some cases.

Anuria is usually due to complete blockage of one ureter, the other kidney being absent, diseased, or similarly blocked. Rarely is anuria the result of blockage of one ureter with reflex suppression of urine in the opposite kidney.

The diagnosis of the causative stone is often suggested by the side on which pain was last felt, the demonstration of renal tenderness and the oedematous and haemorrhagic appearance of the ureteric orifice at cystoscopy. Previous pyelography pictures may be of help but new ones are inadvisable.

With penicillin and chemotherapy to combat infection it is more often possible nowadays to remove the obstructing stone without previous resort to emergency nephrostomy particularly if it is below the pelvic brim, when if necessary both ureters may be explored through the mid-line incision. Above this level accurate determination of the position of the causative stone is necessary in order to ensure its removal; otherwise the safer nephrostomy should be carried out.

At cystoscopy ureteric catheterization is worth attempting in the hope of the catheter passing the obstruction and allowing drainage of urine, the use of antibiotics and chemotherapy being initiated at the same time, so effecting an improvement in the patient's general condition prior to carrying out open removal of the stone.

Forced diuresis with intravenous saline should be effected in all cases and administration of 20 cubic centimetres of 50 per cent sucrose intravenously is of help, but likely to thrombose the vein unless immediately followed by more saline. It must be remembered that although in previously healthy kidneys obstruction may not lead to death from uraemia for 10 days, in these cases the majority of kidneys are diseased and the time before the onset of serious uraemia is much shorter.

9. POST-OPERATIVE CARE

Tropical climate is to be avoided. The stone must be analysed and a suitable diet indicated. In all cases free fluids are advocated, at least a pint of water being given before breakfast.

Diet

Patients with oxalate stones should avoid strawberries, raspberries, rhubarb, currants, tomatoes and spinach; those who have suffered from uric acid stones should avoid entrails, meat soups and wines.

With phosphate stones the urine must be kept acid, and the patient should be taught the use of litmus paper. If Sod. Phosph. Acid., 45 grains thrice daily, is insufficient, Ammon. Nitrate in the form of Stearettes containing 0 5 gramme, 6 being given per day, is a most powerful acidifier.

Chemotherapy

The urine must be rendered sterile by chemotherapy and penicillin. Novarsenobenzol, 0 3 gramme at five-day intervals, is very useful for *Staphylococcus albus* infections.

Stilboestrol, by raising the secretion of citric acid, may be a useful preventative but its effect in causing atrophy of the testes must be considered.

Further x-ray pictures may be recommended for cases likely to relapse.

10. RESULTS OF SURGICAL TREATMENT

The immediate mortality rate in operative treatment is very low, the main complications being paralytic ileus, pulmonary collapse, and secondary haemorrhage when the renal substance has been incised.

Recurrence rate

In assessing late results, infected and sterile cases should be considered separately (Joly, 1929; Winsbury-White, 1946). For infected cases, the recurrence rate may be as high as 30 per cent but with modern chemotherapy, penicillin and novarsenobenzol, this should be much lower in the future.

For unilateral cases requiring nephrectomy, the recurrence rate on the opposite side is low—about 1 per cent. *Bacillus proteus* remains a most difficult organism to eradicate and will render a patient most liable to stone reformation.

Urinary fistula occurs usually because of some obstruction due to stricture or another stone and, fortunately, is surprisingly rare. Similarly, recurrence of a stone in the ureter is rare but may be due to failure to deal with a stricture at the first operation.

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[References to other titles are given under Kidney and Ureter—Stone, in the Index Volume The subject is also dealt with under the heading of Kidney, Surgical Diseases, in the *British Encyclopaedia of Medical Practice* (1938), Vol 7, p. 380]

KIDNEY AND URETER—TUBERCULOSIS

By DAVID BAND, F.R.C.S.Ed.

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1. DEFINITION

213.] Renal tuberculosis occurs as an ulcero-cavernous lesion in the cortex and medulla of the kidney during the stage of visceral dissemination of the disease Symptoms are usually referable to the bladder, and are associated with pyuria and increased frequency of micturition. The tubercle bacillus reaches the kidney *via* the blood stream. Successful implantations of tubercle bacilli may lead to miliary, or relatively few, tubercle follicles in the renal parenchyma. Such foci may be active or quiescent. Nevertheless, quiescent foci may become reactivated after an interval of several years. The presence of renal tuberculosis may be inferred from the detection of tubercle bacilluria,

Blood-borne infection

Tubercle bacilluria

in the absence of pyelographically demonstrable ulcero-cavernous lesions in the kidney. Open renal tuberculosis is always accompanied by the presence of tubercle bacilli in the urine and the characteristic pyelographic changes of ulcero-cavernous lesions *in the kidney. Renal tuberculosis spreads to the* ureters and bladder by implantation of bacilli from the infected urine.

2. INTRODUCTION

Primarily a generalized infection

There is no greater fundamental principle in the consideration of the disease than that tuberculosis is a generalized infection with focal manifestations. In the invasive stage tubercle bacilli infect the lymphatic system. An entrance is gained through the tonsils and the glands in the neck, through the lungs and the peribronchial lymphatics, or through the intestine and the mesenteric lymphatics. The bacilli are air borne or ingested. Once the lymphadenoid tissues have been invaded successfully the centripetal flow of infected lymph carries the organism to the blood stream via the great lymphatic trunks (Munro, 1940). In this manner, implantations of tubercle bacilli from the blood stream reach the viscera, the serous surfaces, the bones and the joints. Their release may be dramatic and overwhelming, as in miliary tuberculosis or tuberculous meningitis; on the other hand, it may be elusive and insidious *as in early visceral tuberculosis of the lung or alimentary tract.*

Stage of visceral spread

3. AETIOLOGY

Whether *the implantations of tubercle bacilli to the tissues are lymphadenoid* or visceral, the nature of the lesions depends on: (1) the number and virulence of the organisms; and (2) the resistance of the host. Accordingly, in the pathogenesis of tuberculosis in any system the existence of a stage at which the lesions are minimal and subclinical must be conceded.

Subclinical stage

The symptoms and signs of renal tuberculosis are those of an open lesion. The clinical syndrome of renal tuberculosis is that of pyuria and increased frequency of micturition, with a characteristic cystoscopic picture (*see* page 317) and the characteristic pyelographic deformity of an ulcero-cavernous lesion in the kidney. Such a cavity in the parenchyma of the kidney is in direct *communication with the calyces into which it has ulcerated.* Tubercle bacilli and pus cells are constantly present in the urine and these should be readily demonstrable in films made of the centrifuged deposit from adequate specimens of urine (Dukes, 1939). Cavity formation in the tuberculous kidney is a visceral lesion quite as gross as that in phthisis, and in order to retrace the stages in its development it occurred to Medlar (1926) to subject to serial section the kidneys removed at necropsy from patients who had died from extra-urinary tuberculosis. As a result of this research it was reported that bilateral microscopic tuberculous lesions were present in the kidneys of the thirty patients examined. Medlar came to the conclusion that many of *the minute scars present in his sections* were due to tuberculous lesions which had healed.

Clinical syndrome

Renal cavitation

Medlar's work

(1) Tubercle bacilluria

Harris (1929) reported a series of *cases of bone and joint tuberculosis* in children, in which there had been a transient tubercle bacilluria without the signs and symptoms of renal tuberculosis (Table I). He concluded that tubercle

Tubercle bacilluria in children

bacilluria signified the presence of a minute, subclinical tuberculous lesion of the kidney, and that such initial or minor renal lesions frequently healed. This statement was a direct challenge to the view long held, that tubercle bacilluria indicated the passage of tubercle bacilli (in a tuberculous subject) through: (a) a perfectly healthy kidney; or (b) a kidney damaged in any way, but not tuberculous; or (c) a kidney damaged by so-called tuberculous nephritis.

TABLE I

REPORTED FIGURES FOR TUBERCLE BACILLURIA

Author	Total cases	Bacilluria	Per cent	Extra-urinary lesion
Harris	110	25	22 7	Bone and joints
Harris	49	4	8 2	Pulmonary
Brown	104	—	10 0	Pulmonary
Hobbs	100	—	6 0	Pulmonary
Dimtza	183	8	4·4	Extrarenal
Deist	31	12	38 7	Pulmonary
Killeuthner	19	3	15·7	Pulmonary
Lotz	13	3	23 0	Pulmonary
Miller	36	12	33·3	Pulmonary
Mack	20	15	75 0	Extrarenal
Band and Munro	174	25	14 4	Pulmonary
Band	*300*	*64*	*21 3*	*Pulmonary*

(2) Histological studies

Tubercle bacilluria in sanatorium patients

The observation that microscopic renal lesions were bilateral in many sanatorium patients with no suspicion of clinical renal tuberculosis, and Harris's report of a 22·7 per cent incidence of tubercle bacilluria, often transient, in children with bone and joint tuberculosis, led to an investigation at Edinburgh University into the incidence of tubercle bacilluria in sanatorium patients who had no clinical symptoms or signs of urinary tuberculosis. Altogether 300 cases of extra-urogenital tuberculosis were examined, and tubercle bacilluria was found in 21·3 per cent (Table II). It was significant that in this series there was a recovery rate of 23·4 per cent in which there was no recurrence of the bacilluria (Table III). On the other hand, the mortality rate was high—59 per cent (Table IV).

TABLE II

SEX INCIDENCE OF TUBERCLE BACILLURIA

Cases examined		Tubercle bacilli found in urine	Per cent
Males	158	20	12 6
Females	142	44	30 9
Total	300	64	21 3

in the absence of pyelographically demonstrable ulcero-cavernous lesions in the kidney. Open renal tuberculosis is always accompanied by the presence of tubercle bacilli in the urine and the characteristic pyelographic changes of ulcero-cavernous lesions in the kidney. Renal tuberculosis spreads to the ureters and bladder by implantation of bacilli from the infected urine.

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TABLE I

REPORTED FIGURES FOR TUBERCLE BACILLURIA

AUTHOR	TOTAL CASES	BACILLURIA	PER CENT	EXTRA-URINARY LESION
Harris	110	25	22·7	Bone and joints
Harris	49	4	8·2	Pulmonary
Brown	104	—	10 0	Pulmonary
Hobbs	100	—	6 0	Pulmonary
Dimtza	183	8	4 4	Extrarenal
Deist	31	12	38 7	Pulmonary
Killeuthner	19	3	15 7	Pulmonary
Lotz	13	3	23 0	Pulmonary
Miller	36	12	33 3	Pulmonary
Mack	20	15	75 0	Extrarenal
Band and Munro	174	25	14 4	Pulmonary
Band	300	64	21·3	Pulmonary

(2) Histological studies

Tubercle bacilluria in sanatorium patients

The observation that microscopic renal lesions were bilateral in many sanatorium patients with no suspicion of clinical renal tuberculosis, and Harris's report of a 22·7 per cent incidence of tubercle bacilluria, often transient, in children with bone and joint tuberculosis, led to an investigation at Edinburgh University into the incidence of tubercle bacilluria in sanatorium patients who had no clinical symptoms or signs of urinary tuberculosis. Altogether 300 cases of extra-urogenital tuberculosis were examined, and tubercle bacilluria was found in 21·3 per cent (Table II) It was significant that in this series there was a recovery rate of 23·4 per cent in which there was no recurrence of the bacilluria (Table III). On the other hand, the mortality rate was high—59 per cent (Table IV).

TABLE II

SEX INCIDENCE OF TUBERCLE BACILLURIA

CASES EXAMINED		TUBERCLE BACILLI FOUND IN URINE	PER CENT
Males	158	20	12 6
Females	142	44	30 9
Total	300	64	21 3

in the absence of pyelographically demonstrable ulcero-cavernous lesions in the kidney. Open renal tuberculosis is always accompanied by the presence of tubercle bacilli in the urine and the characteristic pyelographic changes of ulcero-cavernous lesions in the kidney. Renal tuberculosis spreads to the ureters and bladder by implantation of bacilli from the infected urine.

2. INTRODUCTION

Primarily a generalized infection

There is no greater fundamental principle in the consideration of the disease than that tuberculosis is a generalized infection with focal manifestations In the invasive stage tubercle bacilli infect the lymphatic system. An entrance is gained through the tonsils and the glands in the neck, through the lungs and the peribronchial lymphatics, or through the intestine and the mesenteric lymphatics. The bacilli are air borne or ingested. Once the lymphadenoid tissues have been invaded successfully the centripetal flow of infected lymph carries the organism to the blood stream via the great lymphatic trunks (Munro, 1940) In this manner, implantations of tubercle bacilli from the blood stream reach the viscera, the serous surfaces, the bones and the joints. Their release may be dramatic and overwhelming, as in miliary tuberculosis or tuberculous meningitis; on the other hand, it may be elusive and insidious as in early visceral tuberculosis of the lung or alimentary tract.

Stage of visceral spread

3. AETIOLOGY

Whether the implantations of tubercle bacilli to the tissues are lymphadenoid or visceral, the nature of the lesions depends on: (1) the number and virulence of the organisms; and (2) the resistance of the host. Accordingly, in the pathogenesis of tuberculosis in any system the existence of a stage at which the lesions are minimal and subclinical must be conceded.

Subclinical stage

The symptoms and signs of renal tuberculosis are those of an open lesion The clinical syndrome of renal tuberculosis is that of pyuria and increased frequency of micturition, with a characteristic cystoscopic picture (*see* page 317) and the characteristic pyelographic deformity of an ulcero-cavernous *lesion in the kidney* Such a cavity in the parenchyma *of the kidney* is in direct communication with the calyces into which it has ulcerated. Tubercle bacilli and pus cells are constantly present in the urine and these should be readily demonstrable in films made of the centrifuged deposit from adequate specimens of urine (Dukes, 1939). Cavity formation in the tuberculous kidney is a visceral lesion quite as gross as that in phthisis, and in order to retrace the stages in its development it occurred to Medlar (1926) to subject to serial section the kidneys removed at necropsy from patients who had died from extra-urinary tuberculosis. As a result of this research it was reported that bilateral microscopic tuberculous lesions were present in the kidneys of the thirty patients examined. Medlar came to the conclusion that many of the minute scars present in his sections were due to tuberculous lesions which had healed.

Clinical syndrome

Renal cavitation

Medlar's work

(1) Tubercle bacilluria

Tubercle bacilluria in children

Harris (1929) reported a series of cases of bone and joint tuberculosis in children, in which there had been a transient tubercle bacilluria without the signs and symptoms of renal tuberculosis (Table I). He concluded that tubercle

bacilluria signified the presence of a minute, subclinical tuberculous lesion of the kidney, and that such initial or minor renal lesions frequently healed. This statement was a direct challenge to the view long held, that tubercle bacilluria indicated the passage of tubercle bacilli (in a tuberculous subject) through: (a) a perfectly healthy kidney; or (b) a kidney damaged in any way, but not tuberculous; or (c) a kidney damaged by so-called tuberculous nephritis.

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Cases examined		Tubercle bacilli found in urine	Per cent
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Females	142	44	30 9
Total	300	64	21 3

TABLE III

RECOVERY RATE IN 64 CASES OF TUBERCLE BACILLURIA FOLLOWED-UP FOR FIVE YEARS

Number of cases		Recovery	Per cent
Males	20	5	25 0
Females	44	10	22 7
Total	64	15	23 4

TABLE IV

MORTALITY RATE IN 64 CASES OF TUBERCLE BACILLURIA FOLLOWED-UP FOR FIVE YEARS

Number of cases		Deaths	Per cent
Males	20	13	65 0
Females	44	25	56 8
Total	64	38	59.0

Value of serial sections

The kidneys of 30 patients who had died from extra-urinary tuberculosis were removed at necropsy for histological study. Serial whole sections, numbering 1,000–1,500, were cut from each half kidney, and every fiftieth section was stained and mounted. Such a technique ensured that a comprehensive scrutiny was made of the renal parenchyma and it was significant that bilateral cortical foci of tuberculous disease were found in 24 of the 27 cases of bacilluria examined. In 3 cases in which there had been no bacilluria in life, the kidneys were similarly examined by serial section. Tuberculous follicles were not detected (Table V).

TABLE V

HISTOLOGICAL INVESTIGATION BY SERIAL SECTIONS OF BOTH KIDNEYS FROM PATIENTS WHO DIED FROM EXTRA-UROGENITAL TUBERCULOSIS

	Tuberculous renal lesions found	Tuberculous renal lesions absent	Total
Tubercle bacilluria present	24	3	27
Tubercle bacilluria absent	0	3	3

Pathogenesis of tubercle follicle

The tuberculous lesions were always bilateral. The minute lesions in the kidneys showed every stage in the development of the tubercle follicle. They commonly arose in relation to a glomerulus or in a capillary between tubules close to a glomerulus (Fig. 134(a)). As the lesion progressed central caseation [illegible] sections [illegible] ome

Glomerulus lesion

surrounded by spindle cells. In others the central area had become hyalinized, and the amorphous caseous-like debris had become walled-off by organizing fibrous tissue cells. The follicles were healing. Elsewhere there was scar tissue only.

Healing in the microscopic tubercle follicle

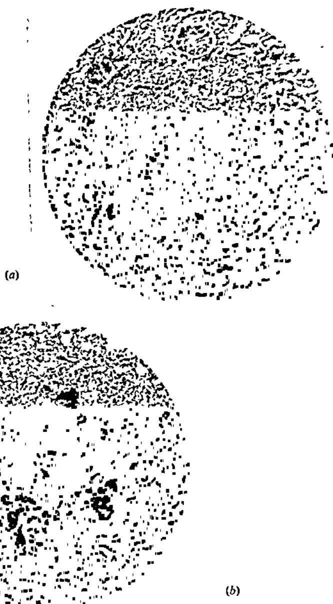


Fig. 134—Tubercle bacilluria (*a*) Typical mononuclear tubercle affecting a glomerulus, (*b*) caseating focus from a field showing giant cells.

Since post-mortem examinations of the kidneys from patients who have died with a positive tubercle bacilluria have shown that microscopic bilateral cortical lesions are constantly present, in all stages of development of the tuberculous follicle, and since 23 4 per cent of patients who have been known to have exhibited tubercle bacilluria without clinical evidence of a

Earliest follicles cortical

renal lesion make a lasting recovery, it may be concluded that: (a) the earliest tuberculous lesions in the kidney are epithelioid and mononuclear tubercles (*see* Plate II (a)). They are situated in the cortex in relation to the glomeruli, and their presence can be detected in both kidneys; (b) the infection is blood borne; and (c) many of such minute lesions undergo spontaneous healing.

(3) Clinical significance

A minute tuberculous focus in the kidney, arising as it does in the glomerulus, may have no connexion with the tubules beyond that of contiguity. Such a lesion may heal and leave behind a minute scar. Under favourable circumstances, such as sparse implantation of bacilli to the kidney, low organismal virulence, and a high degree of resistance on the part of the host, certain lesions which have progressed to the stage of caseation may become walled off, or encysted, by a protective barrier of organized granulations (Fig. 135) Neither healed lesions nor encysted foci discharge tubercle bacilli to the tubules. Encysted lesions, large or small, have no communication with the secretory or drainage system, and are hence termed "closed", and neither pus cells nor tubercle bacilli can be found in urine withdrawn from the kidney. Guinea-pig inoculations are negative. With a heavier or repeated dosage of implanted tubercle bacilli, tuberculous foci in the kidney progress to the stage of caseation and infected debris discharges into neighbouring tubules. The bacilli pass down the tubular system to infect the medulla and the pyramid where contiguous follicles develop at the site of convergence of the collecting tubules. The confluence of a number of such follicles leads to the formation of a readily visible caseating focus at the apex of the pyramid, and, later, to cavity formation with direct communication to the calyces as a result of ulceration (Fig. 136). The open lesion, cortical or medullary, with or without a demonstrable cavity on pyelography, is characterized by the presence of tubercle bacilli in the urine withdrawn from the kidney. Pus cells are constantly present in the centrifuged deposit. But whereas caseo-cavernous renal tuberculosis produces symptoms and signs of a urinary disturbance, and a characteristic pyelographic deformity can be demonstrated in addition to positive bacteriological evidence, the minute cortical lesions which have been seen in the serial whole sections are unassociated with urinary symptoms. Yet, since they are open lesions, there is a tubercle bacilluria, and a few pus cells are constantly present in the urine. When, in a tuberculous subject, there is pyuria without demonstrable cause, the presence of tubercle bacilluria may not be appreciated until cultural and animal inoculation methods have been employed.

Encystment of caseated foci

Closed renal lesions

Formation of the open lesion

Ulcerating open lesion at the pyramid

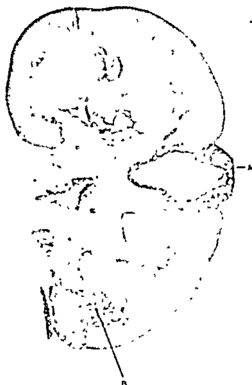
Importance of accurate bacteriological examination

(4) Management of cases of tubercle bacilluria

Tubercle bacilluria signifies the presence in the kidney of minute tuberculous lesions. As the infection is blood borne, these are bilateral. The lesions may be myriad, part of a miliary dissemination; on the other hand, they may be sparsely distributed, and it has been seen that many heal, and that the tubercle bacilluria is sometimes a transient feature (23 4 per cent) Referring to such a stage in renal tuberculosis, Cabot (1933) stated that when a tuberculous lesion in the kidney is closed—that is, when it fails to communicate with the drainage system of the kidney—healing may take place, under favourable conditions. The object of treatment is to permit such minute

Tubercle bacilluria sometimes transient





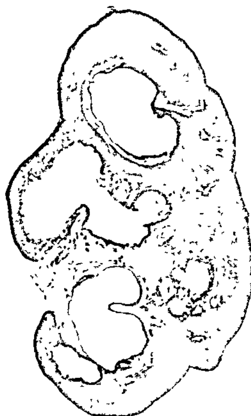
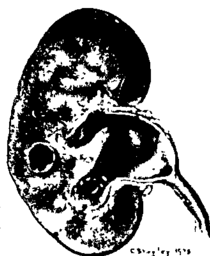
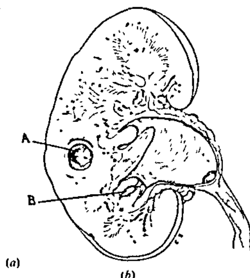


PLATE II

(*a*) Whole section of kidney from case of tubercle bacilluria. Minute foci of tubercle follicles seen at A and B. (*b*) Tuberculous cavity in mid-cortical zone at A, with secondary dissemination, by a process of tubular reabsorption at B, to cortex at lower pole (*c*). Generalized cavitation throughout the kidney, with secondary ulcerative lesions leading to still further dissemination of the tuberculous process



(a)

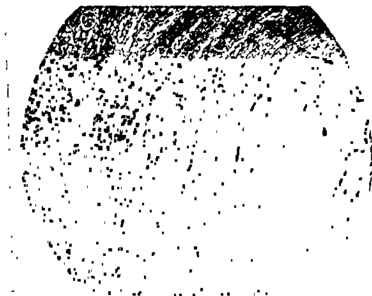


(b)

FIG. 135.—Tubercle bacilli isolated from the urine of a female, aged 22. (a) Drawing of bisected kidney showing an encysted cortico-medullary focus of tuberculosis and cavity formation at the papilla of the pyramid at the lower pole. (b) Diagrammatic drawing of (a). A = Encysted cortico-medullary chronic tuberculous focus. B = Cavitation and calcification in chronic focus at papilla. Tiny tuberculous follicles in adjacent cortex. (c) Microphotograph of epithelioid and mononuclear tuberculous focus in cortex at the lower pole of kidney. (d) Microphotograph of margin of encysted cortico-medullary focus at A in the drawing (b)



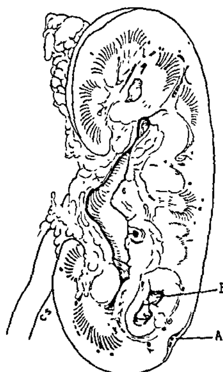
(c)



(d)



(a)



(b)



(c)



FIG. 136 —Tubercle bacilluria in a female, aged 31. (*a*) Drawing of bisected kidney showing depressed scar of the primary cortical lesion now healed and, subjacent to it at the lower pole of the kidney, a thick-walled tuberculous cavity containing calcareous debris. (*b*) Diagrammatic drawing of (*a*). A = Depressed area of primary cortical lesion now healed. *B* = Calcified tuberculous cavity in medulla. (*c*) Tuberculous focus with advanced caseation from area B in (*b*). (*d*) Fibrosing cortical focus at A in (*b*) showing the tendency to heal by fibrosis

lesions as have been diagnosed, on the presumptive evidence of bacilluria, to heal, or to reduce the renal lesions to a frank caseo-cavernous tuberculosis limited to one kidney, which may be extirpated at operation. To attain this, patients who present themselves with tubercle bacilluria must remain under observation until (a) the tubercle bacilluria ceases and the minute microscopic renal follicles, which have caused it, have healed or become encysted; or (b) pyelographic changes of ulcero-cavernous tuberculosis have been demonstrated in one kidney, when surgical intervention becomes necessary; or (c) a general dissemination and miliary tuberculosis render a fatal issue inevitable. During the period of observation the patient should have the benefit of general medical supervision under sanatorium conditions, and extra-urogenital foci should be given appropriate treatment, of which the first principle is general and local rest.

Conservative outlook essential

Period of observation under sanatorium conditions

4. PATHOLOGY OF RENAL TUBERCULOSIS

The stage of ulceration of the renal papilla, and the communicating cavity between the cortico-medullary zone and the renal pelvis, are recognized clinically in the pyelogram. The presence of tubercle bacilli leads to infection of the renal pelvis, and tuberculous ulceration occurs in the lining epithelium. Whereas in the earliest stages there may be a tendency to polyuria and hypermotility, increasing involvement of the pelvic wall leads to atony and stasis which may be enhanced by the presence of tubercles at the pelvi-ureteral junction. By a process of tubular reabsorption, tubercle bacilli in the residual urine of the renal pelvis reach adjacent parts of the renal parenchyma. The organisms pass through the incompetent papillae of the neighbouring calyces, and, by dissemination through the tubules and lymphatics, lead to the formation of secondary tuberculous follicles which by confluence are followed by widespread caseation, ulceration and cavitation throughout the kidney (*see* Plate II (b)). Once a tuberculous lesion in the renal parenchyma has become established and has extended to the tubules, pyramid and papilla, there is a steady progression of the disease throughout the entire organ. The term ulcero-cavernous tuberculosis is applied to the stage of progressive ulceration of neighbouring calyces associated with a primary active focus with cavitation in the parenchyma. The term caseo-cavernous tuberculosis is used when there is a predominance of cavity formation within the kidney (*see* Plate II (c)). Both types may be recognized pyelographically when the shaggy or ragged margin of a widened calyx indicates active tuberculous ulceration; in caseo-cavernous tuberculosis, however, the irregular cavities themselves are the dominant features of the pyelogram.

Ulcero-cavernous lesion

Auto-dissemination by tubular absorption

Ulcero-cavernous lesion

Caseo-cavernous lesion

5. BACTERIOLOGY

The presence of pus cells, without organisms, in an acid urine has for long been the recognized clue to a diagnosis of urinary tuberculosis. Dukes (1939) has recommended the early morning specimen, and a large 24-hour sample of urine as those most suitable for submission to the bacteriologist. Tubercle bacilli should be demonstrable in films made from adequate specimens of urine. Cultural and inoculation methods are necessary, when difficulties in diagnosis occur, or when the type of organism is being investigated (Table VI)

Importance of the 24-hour specimen

TABLE VI

TYPES OF TUBERCLE BACILLUS IN BACILLURIA (SCOTLAND)

HUMAN	BOVINE	TOTAL	BOVINE (PER CENT)
59	5	64	7 8

6. CLINICAL PICTURE

(1) History of the illness

The history of the illness and its insidious onset are important factors Tuberculosis has a familial and social background which has not been overcome by modern developments in social science—for example, infection by the bovine type of tubercle bacillus is still significantly frequent in rural districts in Scotland. A search should be made for the stigmata and symptoms or signs of extra-urogenital foci of tuberculosis, remembering that there may be a considerable interval of years between the invasive stage of tuberculosis in the child, and the stage of visceral spread in the adolescent and young adult.

(2) Symptoms and signs

Frequency

The earliest symptoms are those referable to the bladder. An increased frequency of micturition in a young adult, occurring with pyuria in an acid urine, form a combination of symptoms and signs which are extremely suggestive of tuberculosis. The frequency is constant both by day and by night, and, over a period of months, the increase is gradually progressive.

Haematuria

Renal pain

Haematuria is an inconstant but significant sign in the early stages of renal tuberculosis. It is often transient. Renal pain may occur occasionally; it is usually limited to a dull ache in the loin. Rarely, actual enlargement of the affected kidney may be detected.

Pyuria

Pyuria is constantly present, although it may be no more than an opalescence in the urine which has not been noted by the patient.

7. DIAGNOSIS

(1) Bacteriological

Repeated bacteriological examination

Bacillus coli infections of the urinary tract occur, like those of the *B. tuberculosis*, in an acid urine. However, *B. coli* are readily demonstrable by film or culture. The isolation of the tubercle bacillus is much more difficult. Repeated bacteriological examinations of the urine are essential.

(2) Clinical investigation

Housing conditions

(i) *Social considerations.*—The family and social history of the patient may be of significance. Housing and working conditions, both urban and rural, are factors having an important bearing on the management of the case.

Extra-urogenital lesions

The clinical examination should be comprehensive, having regard to the exposure of other systems to tuberculosis in the stage of visceral spread, and the effects of extra-urogenital lesions on prognosis.

Genital tuberculosis

(ii) *Digital examination.*—Rectal examination may yield evidence of genital foci of tuberculous infection, particularly in the seminal vesicles and the prostate. Irregular thickening, swelling or softening of the pelvic genital

organs in the young adult are most suggestive of tuberculous infection. The testis and epididymis should be palpated. The hard craggy tuberculous lesion at the lower pole of the epididymis is characteristic, and, rarely, the ductus deferens may be felt to be indurated. Cold abscess and sinus formation are diagnostic signs, and confirmatory evidence may be obtained by laboratory examination of scrapings of granulations or aspirated pus. As a rule tuberculosis does not invade the testis till late in the progress of the disease. More serious is a contralateral spread via the ductus deferens to the opposite epididymis.

(3) Coincidence of urinary and genital lesions

Menville and Priestley (1938) found coincident renal lesions in 51·6 per cent of a series of 62 cases of genital tuberculosis in males studied at necropsy. These figures agree with the clinical reports (Hinman, 1938). In genital tuberculosis the incidence of seminal vesiculitis is over 80 per cent (Borthwick, 1946). The occurrence of seminal vesiculitis is of considerable importance because of the sterility so produced in the male. In such circumstances contralateral ligation of the ductus deferens is a logical procedure, in order to prevent infection of the opposite epididymis and testis.

Seminal vesiculitis and sterility

8. DIAGNOSTIC MEASURES

(1) Radiographic examination

Skiagrams should be made of the chest and the abdomen. The straight skiagram may provide evidence of calcareous deposits in the kidney, in cases of chronicity. By taking skiagrams of the abdomen in two planes, anteroposterior and lateral, and during expiration and inspiration, it should be possible to differentiate between extraperitoneal and intraperitoneal lesions.

Importance of the straight skiagram

(2) Cystoscopy

The cystoscopic appearances of vesical tuberculosis have been described (*see* Bladder—Infections, Vol. 2, p. 108) but before carrying out instrumental examinations it is always wise, in the tuberculous subject, to have some knowledge of the activity of the disease, both extra-urogenital and in the urinary tract. Accordingly, a preliminary period of observation in hospital, under conditions of rest, should enable the clinician to have before him such data as the blood sedimentation rate, and a differential white-cell count, as well as other clinical indications of toxaemia, for example, elevation of temperature. Preliminary sedation, and the use of intravenous Pentothal Sodium, or a low spinal anaesthesia, contribute to the ease of the examination when the bladder is of low capacity and irritable. Evidence of ulceration, congestion or small greyish-yellow tubercles, may be seen cystoscopically in the neighbourhood of the affected ureteric orifice. The ulceration tends to spread towards the lateral aspect of the bladder wall In late cases the ureteric orifice may be drawn up and gaping.

Value of anaesthesia

Cystoscopic appearance

When the bladder capacity is greatly reduced and the illumination of the interior of the bladder is hindered by the degree of congestion, the diseased ureteric orifice may be recognized by chromocystoscopy, after the intravenous injection of indigo carmine. The localization of the disease to the affected ureteric orifice and to one half of the bladder wall is a striking feature.

Chromocystoscopy

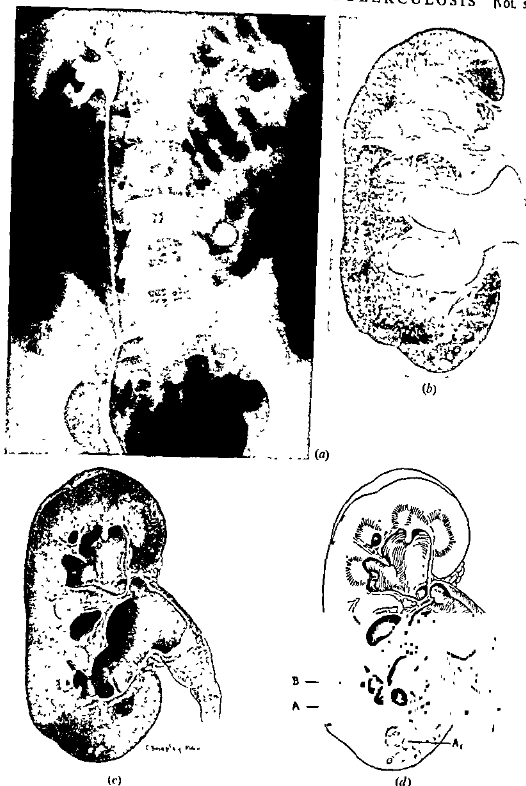


FIG. 137.—Tubercle bacilli present in urine of female, aged 23. (*a*) Pyelogram of the right kidney showing irregularity at lowest calyx (*b*) Whole section of kidney to show follicular character of tubercles at the lower pole (*c*) Drawing of bisected kidney showing active tuberculous follicles at the lower pole with ulceration and cavity formation at the papilla (*d*) Diagrammatic drawing of (*c*) A and A_1: Multiple active tuberculous follicles in cortex. B and B_1: Cavity formation at papilla leading to alterations in pyelographic outline of calyx

When the ureteric orifice is kept under observation it will be noticed that the efflux is cloudy, but not comparable to the tooth-paste-like discharge of a septic pyonephrosis.

(3) Catheterization of the ureters

Intravenous pyelograms are not always sufficiently clearly outlined to be of diagnostic value in the early stages of renal tuberculosis. Emmett and Braasch (1938) insist on the necessity for catheterization of the ureter and retrograde pyelography. A complete examination is essential, and this should include: (a) catheterization of the ureters; (b) bacteriological examination of specimens of urine withdrawn from both renal pelves; and (c) retrograde pyelography. Even if all the information so gained is not necessary for the recognition of tuberculosis of one kidney, a normal pyelogram and a sterile clear urine from the supposedly healthy kidney are clinical observations of profound importance to the surgeon and the patient if nephrectomy is to be undertaken with confidence. Any danger of carrying tubercle bacilli from the bladder to the healthy kidney by regurgitation along the ureteric catheter is unlikely, if over-distension of the bladder and straining are avoided during the examination. Catheterization of the affected ureter may be difficult at the intramural portion, owing to actual stricture formation. As a rule, however, the tuberculous ureter is a thickened and rigid tube which is easily catheterized once the irregular orifice has been negotiated. The urine from the affected kidney is always cloudy, but never offensive. There should be no difficulty in demonstrating pus cells and tubercle bacilli in adequate specimens of urine (Dukes, 1939).

Advantages of retrograde pyelography

Pyelography

When making the pyelogram, over-distension of the renal pelvis should be avoided in order to prevent pyelovenous backflow, which, from a tuberculous kidney, might precipitate miliary spread. The pyelogram is characteristic. The calyx loses its clear-cut, delicate outline (Fig. 137), the cup becomes irregular and the margins become eroded and are often described as shaggy. In the earlier stages intravenous pyelography may lack sufficient clarity of outline to provide diagnostic evidence comparable to that of retrograde pyelography at a similar stage of the disease. As the disease progresses the ulcerative lesion proceeds to cavitation (Figs. 138 and 139) and cavities, which are typically irregular, may be associated with deposits of calcareous debris recognized on the preliminary skiagram. This evidence of chronicity is further exemplified in the reniform shadow of the calcified tuberculous kidney—so-called auto-nephrectomy.

The characteristic pyelographic appearances

9. PROGNOSIS

Untreated tuberculosis of the kidney leads to progressive destruction of the organ, tuberculous ureteritis and cystitis. As a result of ulceration the bladder becomes contracted and tonic. Rarely, the tuberculous ulcers extend across the bladder base to involve the opposite ureteric orifice. Ascending tuberculous infection from the bladder to the remaining healthy kidney is uncommon. More frequently the constant urgency of micturition and the thick-walled contracted bladder produce a backward pressure, which leads to hydro-ureter and hydronephrosis of the remaining kidney, together with

Sequence of pathological changes in urinary tract



FIG. 138.—Renal tuberculosis. Retrograde pyelogram showing early tuberculous cavity at upper pole of right kidney.



FIG. 139.—Pyelogram showing caseo-cavernous renal tuberculosis with gross cavity formation and irregular outline of the ureter.

progressive deterioration in renal function (Fig. 140 (a) and (b)). Death may occur, as a result of toxaemia when there are active extra-urogenital foci of tuberculosis, or from uraemia.

It is only by surgical removal of the tuberculous kidney that cessation of the continued reinfection of the bladder can be brought about, and nephrectomy is the treatment of choice. Fortunately the pathogenesis of the disease, and the latency of visceral spread in tuberculosis, usually present the surgeon with a relatively simple clinical problem. Nevertheless the collaboration of a physician, and a sense of awareness of the quiescence or activity of extra-urogenital foci of tuberculosis should act as a deterrent to simplifying that problem to one of operative surgery alone. Even after nephrectomy and ureterectomy, ulceration in the bladder may persist as a solitary irritable ulcer, close to the roof and lateral wall, which causes persistent frequency and discomfort. Such an ulcer may bleed, or cause a commanding urgency of micturition with attendant loss of sleep and loss of condition, as well as the disablement of invalidism. A contracted bladder may remain after tubercle

Nephrectomy the only curative treatment



(a)

Residual ulceration in the bladder



(b)

FIG. 140—Renal tuberculosis (a) Gross hydro-ureter associated with backward pressure due to the small contracted bladder of chronic tuberculous cystitis, (b) hydronephrosis and hydro-ureter.

bacilli have disappeared from the urine. Months or years after nephrectomy and the eradication of tuberculous foci elsewhere, the persistence of an increased bladder tone may lead to backward pressure on the remaining kidney, hydronephrosis and uraemia. The ultimate mortality rate is between 10 and 15 per cent (Thomson-Walker, 1936; Lett, 1936). The prospect of full recovery with normal frequency, and with rehabilitation to a full and active life as a wage-earner, is under 60 per cent (Band, 1942).

Late uraemia from backward pressure

10. MANAGEMENT

(1) Pre-operative

The consideration of extra-urogenital tuberculous lesions, their state of activity, and their bearing on the condition of the urinary system should be reviewed in collaboration with a physician. Tuberculosis of the kidney is a slowly progressive disease. When the condition has been diagnosed, it may be advantageous to arrange for an interval period of rest in bed under sanatorium conditions prior to operative treatment. If there has been active pulmonary tuberculosis, the physician will decide when the time for operative intervention has arrived. In patients undergoing collapse therapy nephrectomy may safely be undertaken between refillings of the artificial pneumothorax. Although open renal tuberculosis can never be arrested by conservative measures, general rest, attention to diet and the hygiene of a sanatorium regimen all contribute to a marked improvement in the general condition of the patient. This is reflected by a gain in weight, and a lowering of the blood sedimentation rate. In bilateral renal tuberculosis the sanatorium regimen may prolong life with a modicum of comfort for several years, because the tuberculous process, though not arrested, is slowed down, and the gain in the resistance of the patient is reflected in a tendency to calcification in the kidneys, and a diminution in the vesical irritability. Urogenital tuberculosis alone is so slowly progressive that a preliminary period of treatment and observation of a few months' duration may determine: (a) the practicability of operative intervention; (b) a satisfactory post-operative course; and (c) the ultimate successful rehabilitation of the patient after a prolonged convalescence.

Collaboration with physician

Coincident pulmonary tuberculosis

(2) Operative

The only curative therapy for urinary tuberculosis is nephrectomy, in conjunction with the sanatorium life for six months or a year. It is only by surgical removal of the tuberculous kidney that cessation of the continued reinfection of the bladder can be achieved.

Nephrectomy plus post-operative care

(a) Nephrectomy

The operation for removal of the kidney should be carried out extraperitoneally. A gentle technique with an adequate exposure are essential if the risk of dissemination of the disease at the operation is to be reduced to a minimum The perirenal fat should be removed with the kidney, otherwise, when cortical groups of tubercles have infected the extraperitoneal tissues, the forcing of an imperfect line of cleavage between the renal capsule and the surrounding fat may lead to a tuberculous wound infection with sinuses. Pyelovenous dissemination may lead to a miliary spread, and, accordingly, early ligation of the renal pedicle is advisable during the mobilization of the kidney.

Adequate exposure

Early ligation of pedicle

The ureter is a rigid and irregularly thickened tube. It should be mobilized with the immediately adherent peri-ureteric areolar tissues intact. Mobilization beyond the pelvic brim is probably unnecessary. The length of ureter removed should not have been obtained by an operation unduly prolonged, or by an access which required powerful retraction of the wound margins. The employment of two incisions will secure the removal of the entire ureter under vision. The length of the operation time is increased, however, and it is questionable whether the stump of the ureter left behind is of any importance as a source of reinfection of the bladder. Once the kidney has been removed the ureter becomes functionless and the foci of tuberculosis already present in the wall become fibrosed and heal. Reinfection of the bladder ceases when the kidney is removed. In the patient who has recently been toxaemic and in whom extra-urinary foci are present, though quiescent, nephrectomy, along with removal of a length of ureter easily accessible through the incision for exposure of the kidney, is the operation of choice. Similarly it is unnecessary to elaborate the treatment of the divided end of the ureter. The division may be made between clamps, by diathermy or with the carbolized knife. Small strips of packing are inserted round the ureter, and between it and the iliac vessels, prior to the division, to prevent spilling of caseous debris or accidental cauterization beyond the ureter. Thereafter a ligature of chromicized gut is applied and the ureteric stump may be dropped back to the retroperitoneal tissues, with or without a strip of dental rubber for purposes of drainage.

Length of ureter to be removed

(*b*) *Nephrectomy in bilateral renal tuberculosis*

Nephrectomy is usually contra-indicated when bilateral renal lesions are present. Under certain conditions, however, the operation may be undertaken. When tubercle bacilluria occurs alone, without pyelographic evidence of open ulcero-cavernous lesions in the opposite kidney, surgical removal of the more seriously involved kidney may still be curative. The toxaemia induced by the actively diseased kidney may have led to reactivation or to an infection *de novo* of its neighbour which, being minimal and subclinical, may respond to favourable and prolonged sanatorium therapy.

Limited but definite indication

(*c*) *Renal tuberculosis in pregnancy*

Should unilateral renal tuberculosis occur during pregnancy, the kidney may be removed and the pregnancy allowed to go to term. If the lesions are bilateral, however, the pregnancy must be terminated.

11. LATE CONSIDERATIONS

(1) Local treatment of the bladder

The local treatment of the tuberculous bladder after nephrectomy is of less importance than the general management of the post-operative regimen. The persistent residual ulcer may call for local instillations to the bladder of 25 cubic centimetres of a 5 to 10 per cent solution of carbolic acid every fourth day Sometimes, as for interstitial ulcers of the bladder roof, cystoscopic fulguration of the ulcer margin may be valuable. The very irritable bladders may be soothed by the instillation of 25 cubic centimetres of cod-liver oil twice daily. In others a course of tidal lavage with a 1 in 1,000 solution of proflavine or a 1 in 10,000 solution of silver nitrate may reduce the frequency of micturition.

(2) Treatment of persistent contractures of the bladder

Contracted bladder and backward pressure

Reference has been made to the late effects of the contracted and tonic bladder on the intramural portion of the ureter, leading to backward pressure on the remaining kidney. Hydronephrotic atrophy may lead to uraemia and death. There is usually a persistent area of vesical ulceration which fails to respond to treatment. The development of the dilated and tortuous ureter, accompanied by hydronephrosis, may be followed clinically by means of pyelograms. Transplantation of the ureters to the bowel should be carried out while the ureter is still relatively slightly dilated and tests for renal function are satisfactory. Otherwise a cutaneous ureterostomy may yield the patient an expectation of life in fair comfort for many years, provided that the ureterostomy tubes and the collecting apparatus are properly supervised. The cessation of frequency and pain on micturition, and of recurrent haemorrhage from vesical ulcers, and the gain in strength as a result of proper sleep and free urinary drainage, may render cutaneous ureterostomy equally useful when tuberculosis develops in the remaining kidney in the later stages of urinary tuberculosis (Keyes, 1940). Even when considerable dilatation of the ureter has occurred a staged uretero-sigmoid anastomosis may be undertaken by doing a preliminary nephrostomy prior to the actual implantation of the ureter to the bowel. The practicability of uretero-sigmoid anastomosis, even when the ureter is dilated, is undoubtedly the result of preliminary preparation of the bowel by phthalistatin or streptomycin, and the employment of an extraperitoneal approach for the anastomosis. The object of the defunctioning operation on the bladder, whether it takes the form of cutaneous ureterostomy or of uretero-sigmoidostomy, is to bring about healing of the vesical ulcers by local rest to the organ. At the same time free urinary drainage [illegible]

Cutaneous ureterostomy

Uretero-sigmoid anastomosis in stages

Object of defunctioning operations

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LACRIMAL APPARATUS—INJURIES AND DISEASES

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1. ANATOMY

214.] The lacrimal apparatus is made up of two elements: a secretory system and an excretory system. The secretory system is the lacrimal gland which is divided into three parts—a large deeply placed orbital portion lying in the anterior upper and outer aspects of the orbit behind the zygomatic process of the frontal bone, a smaller palpebral portion which bulges into the outer part of the superior fornix, and a series of isolated mucous glands in the superior fornix itself. Fine ducts open into the superior fornix.

Secretory system

The excretory system consists of the two lacrimal puncta, the canaliculi, lacrimal sac and nasolacrimal duct.

Excretory system

The puncta are situated on a slight elevation on the inner ends of each lid, in line with the openings of the Meibomian glands. From these the canaliculi pass vertically for 2 millimetres and then in a more horizontal direction for 8 millimetres where they meet and open into the lacrimal sac either separately or by a common duct.

Puncta
Canaliculi

The lacrimal sac is the slightly dilated upper end of the nasolacrimal duct and it lies in the lacrimal fossa between the anterior and posterior lacrimal crests. It is covered by skin, medial palpebral ligament, orbicularis muscle and a layer of periorbital fascia attached to the anterior lacrimal crest.

Lacrimal sac

The nasolacrimal duct passes down a bony canal formed by the maxilla, lacrimal bone and inferior turbinate to open into the inferior meatus of the nose. Its direction is given by a line joining the inner angle of the palpebral fissure and the first upper molar of the same side.

Nasolacrimal duct

2. PHYSIOLOGY

Function of tears

The function of the tears is to keep the cornea and conjunctiva moist, to assist the action of the orbicularis muscle in sweeping foreign bodies from the corneal surface to the inner corner of the palpebral aperture, and to provide an anti-bacterial lysozyme which inhibits the growth of pathogenic organisms or destroys them.

Passage of tears

Normally just enough lacrimal secretion is provided to keep the cornea and conjunctiva moist and to allow for evaporation, but in conditions of physiological and psychological stimulation, there is an increased flow and the excess passes down the nasolacrimal ducts. This is aided by the capillary action of the canaliculi, the numerous valve-like structures in the nasolacrimal duct and the pumping action of the orbicularis muscle, especially that part of it—Horner's muscle—which passes behind the sac to be inserted into the posterior lacrimal crest. The upper punctum and canaliculus seem to be of little practical value, possibly because in the erect attitude the lacrimal fluid tends to collect in the lower fornix.

3. PATHOLOGICAL CONDITIONS AFFECTING THE SECRETORY SYSTEM

(1) Acute dacryoadenitis

Acute dacryoadenitis is not common. It shows itself as a tender swelling which fills up the interval between the globe and the upper and outer margin of the orbit. It usually settles down, but if fluctuation is detected, the abscess should be drained by an incision just below the orbital margin through the skin, superficial fascia, orbicularis muscle and septum orbitale. The opening into the cavity is enlarged with sinus forceps, the pus is evacuated and a rubber drain inserted for 2 or 3 days.

Treatment

Differential diagnosis.—Orbital cellulitis usually produces a more generalized swelling and a greater degree of proptosis

(2) Chronic dacryoadenitis

Chronic enlargement of the lacrimal gland may be due to chronic inflammation or to new growths.

The chronic inflammatory swellings are often associated with similar swellings in the salivary glands and are included generally in the term Mikulicz's syndrome. They may also be tuberculous or syphilitic. In any case surgical intervention is not usually necessary.

(3) New growths

New growths are mostly slow-growing endotheliomas with a low degree of malignancy in the initial stages; more rarely they are rapidly growing, highly malignant sarcomas

If the gland is steadily increasing in size, removal is advisable.

Treatmen

Under a local, or preferably a general, anaesthetic a curved incision is made from the centre of the superior orbital ridge to the level of the external canthus. Skin, superficial fascia, orbicularis muscle and septum orbitale are incised and the gland is exposed in its capsule. It can be drawn into the wound by blunt dissection and division of fibrous bands attached to the periosteum.

The vessels pass up from below and can be clamped and tied off with catgut or sealed off by coagulation diathermy. If necessary, the palpebral part of the gland lying under the outer part of the levator palpebrae superioris is exposed and removed.

4. PATHOLOGICAL CONDITIONS AFFECTING THE EXCRETORY SYSTEM

Preliminary examination

When a patient complains of excessive lacrimation, a careful inspection should be made of the eyelids for inturned eyelashes, of the conjunctiva for concretions and of the cornea for foreign bodies or ulcers. If this is negative, the excretory system should be investigated.

The puncta are usually visible and should be slightly inverted towards the conjunctiva. There should be no regurgitation of muco-pus into the conjunctival sac when pressure with the finger is applied over the lacrimal sac.

To assess the efficiency of the drainage system, a colouring reagent can be instilled into the conjunctival sac and its appearance noted on the patient's handkerchief when he blows his nose. The passage can also be tested by syringing. A few drops of 1 per cent Pantocaine are instilled and a Nettleship dilator is inserted vertically into the lower punctum with one hand, while the other puts the lower lid on the stretch.

Syringing the nasolacrimal passage

When it is engaged, the dilator is rotated several times and then depressed at right angles, and further rotating movements are made until the punctum is seen to be sufficiently dilated to receive the cannula of a lacrimal syringe. It is then withdrawn and the cannula is inserted vertically and then along the canaliculus into the sac; after a slight withdrawal, the fluid is expressed from the syringe without force. This may pass straight through into the nose, in which case the patient reacts typically to the slight irritation, or it may come out via the upper punctum only. In this case there is probably a block somewhere between the lower end of the sac and the nasal ostium.

If an obstruction is found, it is of the utmost importance to obtain an expert examination of the nose and sinuses. Conditions which may be present are nasal lupus, acute or chronic inflammation in the corresponding antrum, or new growth in the accessory sinuses. Generally, however, the rhinologist reports a perfectly healthy nose.

5. DERANGEMENTS OF THE EXCRETORY SYSTEM

Epiphora

The main symptom is excessive lacrimation, the overflow of tears causing soreness of the skin and giving considerable inconvenience to the patient.

The source of the trouble may be in the puncta, canaliculi or nasolacrimal passage.

(1) The puncta

Due to occlusion of puncta

The puncta may be constricted, especially in the elderly. Dilatation alone may afford temporary relief; more permanent cure is affected by slitting, or by the three-snip operation.

(a) *Slitting operation*

After instillation of a drop of Pantocaine, the punctum is dilated. A canaliculus knife is inserted and pushed gently along the canaliculus. The

handle is then elevated slightly and the medial rim of the fibrous ring is divided for about 1 millimetre. A small plug of cotton-wool is inserted into the canaliculus and left *in situ* for 48 hours. This prevents a too complete repair of the small cut.

(*b*) *Three-snip operation*

A local anaesthetic is injected around the lower canaliculus and the punctum is dilated. A cut is made with sharp scissors through the whole thickness of the conjunctival aspect of the vertical part of the canaliculus. A second cut is made for 5 millimetres along the horizontal portion of the canaliculus, and the triangular piece of tissue is then picked up with forceps and removed by a third snip with the scissors (Fig. 141).

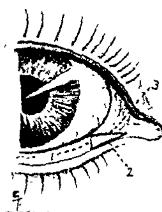


FIG. 141.—Three-snip operation (modified)

Due to ectropion

Eversion of the inferior punctum may be due to a localized ectropion of the inner part of the lower lid, or it may be part of a general ectropion of the whole lower lid.

Treatment by cautery

In the first case replacement can be obtained by a series of cautery punctures deep into the conjunctival surface of the lid just below the punctum. In the second a more extensive plastic operation will be needed.

(2) The canaliculi

Trauma

The canaliculi may be blocked by trauma, inflammation or concretions.

A wound of the lid between the lower punctum and the internal canthus may divide the canaliculus.

Early cases

In the early cases it is sometimes possible to pass a fine probe through the distal, and into the proximal, portion of the duct. The wound is then cleansed and sutured, the probe being left *in situ* for a week and then removed. Repeated probing is necessary afterwards to prevent secondary stricture.

Late cases

In the late cases in which there is continued lacrimation from an impermeable stricture, Stallard's operation can be attempted. In this the sac is isolated and then brought through an incision into the lacus lacrimalis. The sac wall is sutured to the conjunctival edges of the wound and the apex of the fundus of the sac is cut off. There is thus a direct communication between the conjunctival sac and the interior of the lacrimal sac.

Infective stricture

Occasionally after severe conjunctival infections a stricture develops in the canaliculus. This can usually be broken down by passing a probe.

Streptothrix

Concretions are not uncommon in the lower or upper canaliculus. They are caused by the growth of a streptothrix which lodges in the passage. The canaliculus becomes swollen and tender and the punctum is patulous, with a wisp of thick, glairy mucus exuding from it. The punctum is dilated and slit, and the yellowish-green concretions are extruded by pressure. No specific after-treatment is required.

(3) The nasolacrimal duct

Obstruction at birth

This may be blocked at birth, and the condition is noticed when lacrimation starts at the second or third week. It is usually unilateral and does not respond

to any form of local treatment. There may be a discharge of muco-purulent fluid from the puncta on digital pressure over the lacrimal sac. The majority of these cases recover naturally during the first 6 months of life, but if the condition persists or if there is much discharge, the obstruction must be overcome.

Syringing Syringing alone may dislodge a plug of mucus, but generally a probe must be passed to perforate the membrane which may be present at the lower end.

Probing In this case a general anaesthetic is given, the upper punctum is dilated and a fine probe is passed down the canaliculus into the sac and then along the course of the nasolacrimal canal until it strikes the floor of the nasal cavity. It should pass without any appreciable sense of resistance. There is no need to syringe the duct for evidence of success, since relief is immediate if the probing has been correctly performed. The upper canaliculus is employed because it is in an almost direct line with the nasolacrimal duct and so any damage to the much more important lower canaliculus is avoided.

Stricture in adults In adults the stricture may be either traumatic or inflammatory. In the former case, there is often much damage to the bony walls of the nose, and treatment is complicated in consequence. In the latter, the stricture is probably inflammatory and is situated midway between the lower end of the sac and the nasal opening. It is a curious fact that 80 per cent of the patients are females and no satisfactory explanation of this high sex incidence has yet been found. It may be connected with the periodic monthly congestion which affects the nasal mucous membrane in the female. The sac may be normal or it may be dilated and infected, with regurgitation of mucus on pressure.

Graduated probing may be tried, but it is a painful procedure unless a local anaesthetic is injected deeply around the duct. Relief is unusual and merely temporary.

(4) The lacrimal sac

This is not so much a sac as the slightly dilated upper part of the nasolacrimal duct which does not lie in a bony canal.

Chronic dacryocystitis A block in the canal below may result in the accumulation of debris in the sac, with secondary infection. This infection spreads to the mucous membrane and walls of the sac, causing thickening or distension, and muco-pus can often be expressed by pressure over the internal palpebral ligament. A not uncommon complication is perforation of the sac with resultant cellulitis. A tense, red, extremely tender swelling appears, which usually needs incision under a general anaesthetic. No damage should be done to the sac itself, for this may be needed later if a curative operation is required The wound usually heals but a fistula may develop. Relapses are common. *Acute dacryocystitis*

In cases of impermeable stricture with little, or even with considerable, chronic infection of the sac, the alternatives of complete removal of the sac or of a restorative operation must be considered. In the first, the infected sac is removed, and with it the danger of infection of the eye if it is injured, but a varying degree of epiphora remains. This is the operation of choice in old people or in emergency operations on the globe when an infected sac must be rapidly eliminated.

(a) *Dacryocystectomy*

Dacryocystectomy is performed under a local or preferably a general anaesthetic. If the former is used, the local anaesthetic solution is first infiltrated

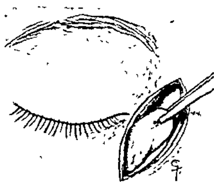


FIG 142.—Dacryocystectomy

into the skin and then, with the needle at right angles to the surface, a deep injection is made above and below the internal palpebral ligament down to the periosteum in the lacrimal fossa.

A curved incision is made from just above the internal palpebral ligament, curving outwards along the anterior lacrimal crest to the lower orbital margin. The orbicularis muscle is divided and a lacrimal retractor is inserted. The only vessel which may give trouble is the angular vein. The anterior lacrimal crest is identified and the internal palpebral ligament divided. This exposes the fascia covering the sac, which is opened. The sac is separated from its bed partly by blunt dissection and partly by means of a small triangular Hudson's knife, starting on the medial side and working over the fundus to the lateral aspect. It is then lifted out and the nasolacrimal duct is severed as low as possible with knife or scissors. A curette is then passed into the bony canal and the mucous membrane is scraped away. The cavity is cleaned out and the skin sutured, the internal palpebral ligament being included in one of the skin sutures; a firm pad is applied to the wound to prevent bleeding (Fig. 142).

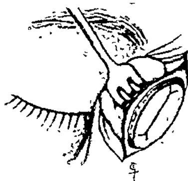


FIG. 143.—Dacryocystorhinostomy. Exposure of sac and nasal mucous membrane.

(*b*) *Dacryocystorhinostomy*

In the reconstructive operation of dacryocystorhinostomy (Figs. 143, 144 and 145), the aim is to create a new passage by anastomosing the mucous membrane of the sac with that lining the lateral wall of the nasal cavity. The approach is identical with that for removal of the sac but, if a local

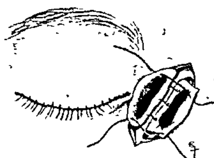


FIG 144.—Dacryocystorhinostomy. Union of posterior flaps.

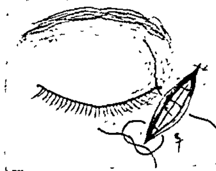


FIG. 145.—Dacryocystorhinostomy. Final sutures in anterior flaps and skin.

anaesthetic is used, some ribbon gauze soaked in 5 per cent cocaine with adrenaline is inserted into the nasal cavity beforehand. The sac is exposed and drawn laterally out of the lacrimal fossa, care being taken not to damage the fundus into which the canaliculi open. The bony floor is removed over an area bounded by the anterior lacrimal crest in front, the lower edge of the internal palpebral ligament above, the posterior lacrimal crest behind and the lower end of the bony lacrimal passage below. Hammer and chisel or a dental burr may be used to make the initial holes, which are then enlarged with punch forceps. The nasal mucous membrane must not be damaged. The sac is released and a probe passed via the inferior canaliculus to check its exact position. Anterior and posterior flaps are then cut in the nasal mucous membrane and through the whole thickness of the sac wall, and silk sutures are inserted into these, uniting the posterior flaps first and then the anterior ones. The wound is closed with interrupted sutures and no after-treatment is needed.

Complications

The complications which, though not altering the technique, may increase the difficulties of the operation are a contracted thick-walled sac with a very small cavity, or adhesions round the sac with possibly a fistula which should be cut out and removed.

The exposure of anterior ethmoidal cells under the lacrimal fossa means an increase in the depth of the field of operation, for these must be cleared away before the nasal mucous membrane can be identified.

Dacryocystorhinostomy can be performed by the nasal route (West's operation) but the external approach gives better access and on the whole the results are more satisfactory.

(5) Removal of palpebral portion of lacrimal gland

In cases of obstinate epiphora, in which there is no possibility of restoring the channel for tears into the nose, the palpebral portion of the lacrimal gland can be removed to reduce the production of tears.

The upper lid is everted and a fixation suture is inserted into the upper edge of the tarsal plate, which is then retracted upwards. An incision is made in the exposed conjunctiva of the outer aspect of the superior fornix, and the gland, which is easily recognized, is dissected out and removed. Many of the ducts from the main portion of the gland pass through this accessory portion; they are thus obliterated and relief is obtained.

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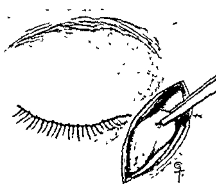


FIG 142.—Dacryocystectomy

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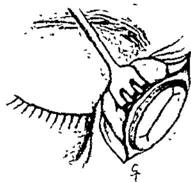


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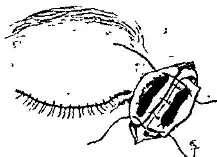


FIG 144.—Dacryocystorhinostomy Union of posterior flaps.

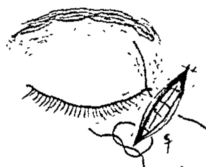


FIG. 145.—Dacryocystorhinostomy Final sutures in anterior flaps and skin.

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(5) Removal of palpebral portion of lacrimal gland

In cases of obstinate epiphora, in which there is no possibility of restoring the channel for tears into the nose, the palpebral portion of the lacrimal gland can be removed to reduce the production of tears

The upper lid is everted and a fixation suture is inserted into the upper edge of the tarsal plate, which is then retracted upwards. An incision is made in the exposed conjunctiva of the outer aspect of the superior fornix, and the gland, which is easily recognized, is dissected out and removed. Many of the ducts from the main portion of the gland pass through this accessory portion; they are thus obliterated and relief is obtained.

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LARYNX—DIRECT LARYNGOSCOPY AND ASPIRATION TREATMENT IN LARYNGEAL DIPHTHERIA

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1. DEFINITION

215.] Laryngeal diphtheria may be primary, the larynx alone being affected, or it may be secondary to faucial or nasal diphtheria or to both.

2. AETIOLOGY

Age

Laryngeal diphtheria is most common between the ages of 12 months and 5 years but it also occurs in older children and, occasionally, in adults.

Bacteriology

The causative organism is the *Corynebacterium diphtheriae* or Klebs-Loffler bacillus which remains localized at the site of infection and produces a powerful exotoxin. The diffusible toxin is absorbed into the blood and is responsible for the various paralyses and for heart failure.

3. PATHOLOGY

The *Coryn. diphtheriae* causes a fibrinous exudation or "membrane", with degeneration, necrosis and desquamation of the superficial epithelial cells. The organisms are found on the surface of the exudate with layers of fibrin [illegible] and bronchi [illegible]

often of a whiter colour than is the membrane on the fauces. It is more [illegible]ent to the true vocal cords and epiglottis where the epithelium is stratified. As a general rule in primary laryngeal diphtheria toxaemia is not marked and late heart failure and paralysis are rare. In secondary laryngeal diphtheria with extensive faucial or nasal membrane, toxaemia is often severe. The degree of toxaemia varies according to the site and extent of the local lesion, but there is considerably less absorption of toxin from membrane in the larynx

than from lesions in the throat and nose. In some cases the membrane may spread downwards, involving the trachea, bronchi and smaller bronchioles—tracheo-bronchial diphtheria. In this dangerous and fatal form of diphtheria, the membrane may involve the whole bronchial system, with considerable purulent secretion in the bronchi. Emphysema follows, and petechial haemorrhages are often found on the surface of the lungs, with extensive areas of collapse and broncho-pneumonia. Degenerative and toxic changes are present in the heart, liver, kidneys and suprarenal glands.

4. CLINICAL PICTURE

Three stages are recognized according to the development of the disease.

(i) *Initial stage.*—The onset is usually gradual and the symptoms are cough and hoarseness with or without moderate pyrexia.

(ii) *Spasmodic stage.*—Aphonia develops with recurrent attacks of inspiratory stridor which increase in severity. The cough is croupy with a high-pitched metallic ring. The child becomes anxious and restless and the face may show slight cyanosis. Inability to inspire enough air to expand the lungs fully leads to recession of the epigastrium and lower intercostal spaces. Restlessness becomes more marked and continuous. Occasionally there is slight but temporary relief if small pieces of membrane or tenacious mucus are coughed up, though the patient is never entirely free from symptoms. Obstructive signs may be accentuated by spasm of the laryngeal muscles. Death may occur in this stage from obstruction of the glottis by a semi-detached fragment of membrane or by thick tenacious mucus. Dyspnoea becomes continuous and the respiratory movements are more violent, with marked recession of the epigastrium, intercostal spaces, suprasternal fossae and subclavicular spaces. A livid hue due to insufficient oxygenation develops and the pulse is small, rapid and often irregular.

(iii) *Permanent obstruction.*—The child is completely prostrated and too weak to struggle. The face is pale and livid; the lips are purple. Restlessness ceases, the respiratory movements become feebler and, if the case is untreated, death supervenes.

5. DIAGNOSIS

Although the existence of laryngeal symptoms with exudate on the fauces or in the nose should always suggest diphtheria, croup with obstructive signs may arise from inflammation caused by other pathogenic organisms. In the absence of faucial or nasal membrane the recognition of laryngeal diphtheria is impossible unless use is made of the laryngoscope In addition direct laryngoscopy is of value in deciding whether operative measures are required, and what should be done. Gover (1918), reporting on 189 cases of "croup" at the Willard Parker Hospital, stated there was no other method nearly so accurate in the diagnosis of laryngeal diphtheria. In 112 of his cases with membrane in the larynx, 53 (47 32 per cent) had no membrane in the fauces.

6. DIRECT LARYNGOSCOPY

The apparatus used consists of Chevalier Jackson's laryngoscopes, infant and child sizes Light is supplied by connecting the lamp carrier by flexible cable *Apparatus*

either to a lighting plug or to a dry-cell battery with a rheostat. Swab-sticks about $9\frac{1}{2}$ inches long and a tongue depressor are required.

Method

An anaesthetic is not required, but the patient must be under perfect control. This is attained by placing the arms downwards in a straight line with the body and wrapping the patient up in a blanket which is firmly secured by large safety-pins. The patient is then placed upon the operation table in the recumbent position. A sand-bag about 4 inches thick is placed under the shoulders and the head is extended far back. The Sister stands on the right of the patient and holds the head firmly and steadily in the midline by a hand placed on each side. Another nurse, standing on the left of the patient, steadies any movements of the limbs and trunk. The operator, wearing a face mask and plain spectacles, stands at the head of the table, opens the mouth with a tongue depressor and, holding the laryngoscope in the left hand, inserts it over the tongue and, keeping to the midline, hooks back the epiglottis with the tip of the instrument. The laryngoscope is now pushed farther down and the upper larynx, vocal cords and subglottic region are in view (Fig. 147)

The important points to note are:

(1) the character and distribution of the membrane;
(2) the intensity and situation of inflammation, if present;
(3) the presence of oedema, especially subglottic;
(4) the presence of muco-pus and secretion;
(5) signs of ulceration;
(6) the presence of a foreign body.

Bacteriological examination

A swab is taken by introducing a swab-stick through the lumen of the laryngoscope and applying it gently to the upper larynx and subglottic region. Suitable media (Löffler's blood serum, tellurite and blood agar) are inoculated and a bacteriological report can be made in 18–24 hours.

7. PROGNOSIS

The younger the patient, the more serious is the disease. The outlook is good if laryngeal obstruction can be relieved, provided that there are no extensive faucial or nasal lesions. Broncho-pneumonia, which may follow tracheotomy, is very rare in cases successfully treated by aspiration.

8. INDICATIONS FOR ASPIRATION

Formerly it was customary in hospital practice to wait as long as was reasonably possible in the hope that the patient might cough up the obstructing membrane, and that intravenous antitoxin might prevent further spread The writer is of the opinion that direct laryngoscopy should be carried out as soon as possible, and that aspiration should be performed if obstructive symptoms are due to membrane or to tenacious muco-purulent secretion.

9. TREATMENT BY ASPIRATION

Respiratory distress in laryngeal diphtheria may result from:

Causes of dyspnoea

(1) membrane blocking the larynx, or membrane partially detached and drawn into the lumen of the larynx by inspiration;
(2) inflammation and oedema in the subglottic region;

(3) thick muco-purulent material which the patient is often unable to cough up;

(4) spasm of the glottis.

In 1918, Gover reported respiratory relief obtained in 23 out of 29 patients, by removal of loose membrane from the larynx by forceps. In 1922, in a series of 53 cases of laryngeal diphtheria, Thomson reported good results obtained by removing loose membrane and mucus, using a straight applicator with a screw clasp on the tip, to which a small gauze plug was attached. Under direct laryngoscopy, this was passed into the larynx below the cords and immediately withdrawn. This treatment, repeated on two or three occasions, gave much relief, and avoided the necessity for intubation in 37 cases.

Historical

In 1923, Gover and Hardman, of the Willard Parker Hospital, New York, reported encouraging results in the treatment of 50 cases of laryngeal diphtheria. Gover preferred to use metal tubes whereas Hardman used silk elastic catheters. These were passed into the larynx through the lumen of the laryngoscope and suction was performed by using an electric pump. This procedure lessened the number of cases requiring intubation and so reduced mortality. It is interesting to note that suction was performed 12 times in one case and 10 times in another. In the same year, Litchfield and Hardman, of the Willard Parker Hospital, in a series of 106 cases of laryngeal diphtheria, employed visually guided suction and found that the mortality was reduced.

In 1930, Tolle reported a series of 344 cases of croup, of which 212 were diphtheritic and 132 non-diphtheritic. In patients treated by suction, only 19·8 per cent needed intubation as compared with 41 per cent without suction.

In 1931, Benson reported on the treatment by aspiration of 37 patients with moderate or severe croup and stated that fewer cases of laryngeal diphtheria required operation if aspiration was judiciously employed.

In a series of 55 cases of laryngeal diphtheria treated by aspiration in 1931 and 1932 (reported in 1933), Tregelles found that operation was probably obviated in 16. Her figures tended to support those of the American workers in showing that the incidence of operation for laryngeal diphtheria was lower if aspiration were performed, and she was also inclined to agree with Tolle that the death rate following operation was lower.

In 1934, Lemariey and Hamon reported favourably on 4 years' experience of the aspiration treatment of laryngeal diphtheria in Paris. Their technique is the same as that generally practised, except that they prefer a suction cannula with a blind end, and one or two lateral fenestrae near the tip instead of one with a terminal opening, and in addition to clearing out the larynx and trachea, they apply suction to the main bronchi when necessary.

Apparatus

The apparatus consists of an Atmos suction pump with wash-bottle and three metal laryngeal suction tubes (Willard Parker Hospital model) with terminal aperture and an external diameter of 5 millimetres, 6 millimetres and 7 millimetres respectively. Stout rubber tubing, a basin of sterile water and sterile liquid paraffin are also required (Fig. 146).

Method

Sufficient water is poured into the wash-bottle to cover the end of the inlet tube. The air-tight lid is screwed down and the exit tube attached to the suction pump. The inlet tube is connected to the laryngeal suction tube by stout rubber tubing. Sterile water is drawn through the suction tube to make certain that the apparatus is in working order. The laryngoscope is now

introduced and when the cords are in view, the laryngeal suction tube, lubricated with sterile paraffin, is inserted through the lumen of the laryngoscope, and membrane is aspirated from the upper aperture of the larynx (Fig 147). When glottic spasm relaxes, the suction tube is passed between the true cords, and membrane and secretion are aspirated from the subglottic region. The suction tube is then withdrawn and, after drawing sterile water through the lumen, it is reintroduced well down into the trachea. Although it may be necessary to introduce the suction tube two or three times, the operation can be carried out very quickly without anaesthesia. Separation of the membrane, which is frequently loose and flapping, seldom gives rise to haemorrhage (Fig. 148). One operation does not always give permanent relief and frequently it has to be repeated. Patients with extensive and re-forming membrane usually require aspiration at intervals of 8–12 hours during the first 24–48 hours in hospital.

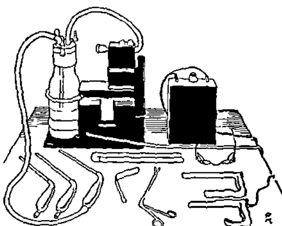


FIG. 146.—Apparatus for direct laryngoscopy and aspiration.

Contra-indication to aspiration

If there is an accompanying or a following subglottic oedema, tracheotomy or intubation may be required. In cases in which subglottic oedema is marked and is causing severe obstruction, aspiration is harmful. Tracheotomy is the method of choice in these cases.

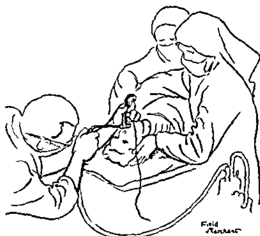


FIG. 147.—Direct laryngoscopy and aspiration



FIG. 148.—Casts of diphtheritic membrane removed from larynx and trachea by aspiration.

10. ADMINISTRATION OF ANTITOXIN

It is essential that diphtheritic antitoxin should be administered intravenously in adequate dosage as soon as possible. Dosage will vary according to the severity of the case, from 20,000 units in primary laryngeal diphtheria to 80,000 or 100,000 units in severe cases with extensive faucial, pharyngeal or nasal membrane.

11. PENICILLIN THERAPY AND CHEMOTHERAPY

In the writer's experience chemotherapy is disappointing but the administration of penicillin—50,000 units intramuscularly at 3-hourly intervals—should be commenced as soon as possible.

12. TRACHEO-BRONCHIAL DIPHTHERIA

When diphtheritic membrane involves the bronchi, the outlook is extremely grave. Aspiration gives no relief. Lynah (1916) found intubation and tracheotomy equally unsuccessful in the treatment of these cases and reported a recovery of 64 per cent of cases following bronchoscopy with suction for the removal of the obstructing membrane. Welford (1929) also found intubation and tracheotomy equally unsuccessful; he recommends large doses of antitoxin (40,000 to 50,000 units) with bronchoscopy and aspiration, if performed before the onset of myocarditis, as the only chance of combating this type of obstruction. In his series of 24 cases, there was only one recovery.

Intubation

The employment of the laryngoscope enables intubation to be carried out with safety and ease by using a suitable introducer. The operation is much simpler than the older O'Dwyer method.

13. CONCLUSIONS

The conclusions to be drawn from the observations made are summarized below.

(1) Direct laryngoscopy is essential for the diagnosis of primary laryngeal diphtheria.

(2) Direct laryngoscopy is valuable in determining the best line of operative treatment to adopt in severe cases of croup.

(3) Cases of croup with obstructive symptoms, which are due to diphtheritic membrane in the upper larynx and subglottic region, and which show little or no subglottic oedema, are relieved by aspiration, and in many cases will be saved the necessity for operation.

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LARYNX—SURGICAL DISEASES OF

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1. FOREIGN BODIES

216.] Impaction of foreign bodies in the larynx is rare. The majority of such objects which enter the larynx are quickly expelled, or if small enough they

may be drawn down into the trachea and bronchi. The exceptional cases in which impaction occurs are those involving objects of irregular shape—such as collar studs—or those having one or more sharp points, such as a safety-pin or a small dental bridge. Portions of such objects may become involved in the laryngeal sinus, or a sharp projection may be caught in one of the folds of the larynx, when movement either upwards or downwards becomes impossible.

Symptoms The impaction of such a foreign body is accompanied by an attack of coughing which will be followed by hoarseness and pain and, perhaps, by some dyspnoea. If the impaction continues, an inflammatory process with oedematous swelling may occur and dyspnoea may become urgent.

Diagnosis Diagnosis can usually be made on inspection by indirect laryngoscopy, but in some cases, particularly when time has elapsed and when inflammatory swelling has occurred, direct laryngoscopy under cocaine and adrenaline anaesthesia will be necessary. Radiography will be of great assistance in the case of metallic objects, but the complex shadow thrown by the laryngeal cartilages, often partly ossified, will reduce the value of x-ray examination in the case of non-metallic objects.

Extraction The foreign body can, in the majority of cases, be removed by forceps through an endoscopic tube. This should be done under local anaesthesia, because of the danger of spasm and asphyxia, or because of the danger of the inhalation of the object into the lower air passages under general anaesthesia. The application of cocaine and adrenaline, perhaps more than of any other local anaesthetic, causes a shrinking of the swollen tissues and facilitates extraction. *Dyspnoea* Obstruction may cause such dyspnoea as to render tracheotomy or laryngotomy necessary before removal is attempted.

Occasionally, extremely urgent dyspnoea is caused by the impaction of a large piece of meat or other similar material which is drawn into the larynx by some sudden inhalation, such as a cough, whilst swallowing A soft foreign body of this nature may completely fill the glottis and produce such obstruction that, unless it can be dislodged, or a tracheotomy or laryngotomy performed within a period which can be measured in seconds, death from asphyxia will occur.

2. INJURIES TO THE LARYNX

Injuries to the larynx may be divided into two groups: those in which the traumatic influence is from outside and those in which the injury occurs inside the lumen.

(1) Injuries due to external influence

(a) Impact with blunt objects

Examples of this type of injury are impact with portions of machinery in motion, the rim of a steering wheel or other object in motor-car accidents, collision with a horizontal wire or railing, a sudden tightening of a rope in suicidal hanging and many other similar occurrences.

The thyroid cartilage and, less commonly, the cricoid cartilage are fractured and the larynx is compressed, causing narrowing of the lumen. Extravasation of blood into the tissues will cause further narrowing and dyspnoea may become severe.

(b) Penetrating injuries

These may be caused by gunshot wounds, by daggers, and by pointed pieces of metal in motor-car and other accidents. Retention of the penetrating object would, in most cases, produce fatal dyspnoea, but even after the exit of a small missile or the removal of a larger one, extravasation of blood and swelling will cause severe dyspnoea.

(c) Incised wounds

Usually, these wounds are due to the suicidal or homicidal use of a razor or knife, and the laryngeal cartilages are divided horizontally. If, as usually happens, the lumen is opened, considerable bleeding will take place and, as the normal protective mechanism ceases to be effective, blood finds its way into the bronchi and lungs.

Bleeding

(d) Diagnosis

The diagnosis of injuries due to external influence will be helped by knowledge of the causative force. It will be confirmed by the coughing up of blood-stained saliva, or by surgical emphysema in the neck, especially in the absence of an external wound. The position and extent of penetrating and incised wounds will give valuable clues as to the probable implication of the larynx. Fractures of the cartilages may be diagnosed by alteration in shape, by undue mobility of parts of the laryngeal skeleton, and by the presence of crepitus on palpation. In many cases bruising and swelling of the superficial soft tissues may make the signs difficult to detect. Radiography will occasionally give assistance in diagnosis, but such evidence will often be inconclusive. Indirect and direct laryngoscopy will give valuable evidence, and the latter will enable blood-stained secretions to be aspirated.

Complications

The danger of these injuries comprises (1) dyspnoea, which may be due to compression, to bleeding into the air passages or to surgical emphysema of the neck which may spread to the mediastinum, and (2) the general effects of haemorrhage, which may be severe. If the pharynx or oesophagus is also involved the danger of infection may be added.

(e) Treatment

The treatment of the injuries described depends upon whether the air channel is narrowed enough to cause embarrassment, whether there is active haemorrhage or whether there is surgical emphysema.

In the absence of these complications it is wise to keep the patient quiet in bed and to avoid the use of opiates and of atropine, as they dry up secretion and aggravate or precipitate dyspnoea.

Reduction of fractures

Bruising and swelling of the soft tissues may be reduced by the application of cold compresses. Slight palpable displacement of portions of the cartilages may be manipulated from outside, and kept in position by a moulded stent splint. Intralaryngeal manipulation by means of endoscopy may be necessary to replace depressed portions of the cartilage.

Intralaryngeal splinting

It may be difficult to keep such portions in place, unless a mould of stent or of gauze be introduced and fixed inside the larynx, after a tracheotomy has been performed. This mould must be kept in place for at least 7–10 days, until the lumen keeps open independently when the cartilages have become fixed in the proper position. It is much better to carry out this reduction as soon a

the extravasation and swelling inside the larynx have subsided than to allow the deformity and stenosis to become established, when either a laryngofissure and skin graft would be necessary (*see* page 361) or a tracheal cannula would have to be worn indefinitely.

In penetrating wounds

Tracheotomy.—A temporary tracheotomy has often to be performed in cases of penetrating wounds because of the swelling of the soft tissues in the laryngeal interior, but can be dispensed with after a few days when the swelling has subsided. Except for attention to oral hygiene and toilet to the external wounds other treatment is not usually necessary, but varying degrees of hoarseness may subsequently call for speech therapy and scarring may call for plastic measures.

In incised wounds

For incised wounds of the larynx, tracheotomy may often be necessary because of the amount of haemorrhage. The tracheal opening enables blood in the trachea and bronchi to be aspirated and, if the bleeding is persistent, it allows a gauze pack to be inserted into the laryngeal lumen by means of direct endoscopy.

It is desirable that, after toilet and the application of penicillin and a sulphonamide powder, the wound be closed so far as is possible, but the retention of the tracheal cannula for a few days is advisable until the danger of surgical emphysema has passed. An attempt should not be made to anchor the divided fragments of a cartilage to each other directly as this will only cause necrosis of portions of the cartilage, but sutures may be placed in the perichondrium and attached muscles.

(2) Injuries occurring inside the laryngeal lumen

Causes

These may be due to impacted foreign bodies, to the passage of endoscopic tubes and instruments, to intubation either of O'Dwyer's tubes or of intratracheal anaesthetic cannulae, or to the effects of too high a tracheotomy. Injury may also be caused by misdirection of a needle in the introduction of radio-opaque oils through the cricothyroid membrane into the trachea. Such

Symptoms and treatment

injuries are usually comparatively slight, cause temporary hoarseness, or aphonia, some bleeding, and occasionally dyspnoea which may require temporary tracheotomy. Usually rest and silence, together with the dry inhalation, by means of a Burney Yeo perforated metal respirator or similar appliance, of a volatile substance such as menthol, eucalyptus oil, Chloretone or creosote will cause healing in a few days.

Sequelae

An occasional sequela may be the formation of a semi-pedunculated granuloma at the posterior end of one of the vocal folds and this can readily be removed by endoscopy. Extensive scarring may occur after injury by a very high tracheotomy and may require plastic measures as described on page 365.

3. INFLAMMATORY DISEASES

(1) Acute laryngitis

Aetiology

Acute infection of the larynx is a very common condition which is associated with the common cold, influenza, the acute exanthems, exposure to cold and wet, and over-use or misuse of the voice. In many cases there is a concurrent

Symptoms

acute pharyngitis. The symptoms are hoarseness, pain, dysphagia and pyrexia, but these can be treated by medicinal measures such as gargles, sprays and inhalations. The common acute laryngitis does not, as a rule, call for surgical

measures but there is a fulminating type which is characterized by oedema and in which dyspnoea is present or imminent. This oedematous form is caused by the presence of a virulent type of haemolytic streptococcus, and is associated with an acute pharyngitis or tonsillitis. It may also be associated with peritonsillar abscess or quinsy, and with Ludwig's angina and other acute infections of the floor of the mouth and tongue. It is frequently present after swallowing corrosive fluids, or after scalds from drinking very hot fluids It occurs occasionally in typhoid fever and frequently in agranulocytic angina.

Oedematous form

The mucosa of the epiglottis, aryepiglottic folds and arytenoids, and even of the vestibular folds, becomes enormously swollen and asphyxia may become imminent. Hoarseness and some pain are early symptoms.

In the past, these cases gave rise to the greatest anxiety and at times required tracheotomy, but by the use, first of anti-scarlatinal serum and later of the sulphonamides, the oedema due to the infectious laryngitis could be rapidly controlled. An initial injection of 30 cubic centimetres of the serum will often avert a catastrophe and the full application of a sulphonamide—8 grammes within the first 24 hours followed by 4-hourly doses of 1 gramme—has an even surer effect. Massive doses of penicillin are equally efficacious. Morphine, Omnopon or other narcotics, and atropine should never be given as they dry up the mucous secretions and increase dyspnoea. Similarly intralaryngeal manipulations should be avoided as they are likely only to increase the swelling and inflammation.

Treatment

In the cases due to corrosive fluids and scalding, serum and sulphonamides have much less effect, and after the immediate application of the proper antidote the sucking of pieces of ice and the application of iced compresses to the neck may avert dyspnoea and the need for tracheotomy.

Tracheotomy

Although some hesitation in performing tracheotomy in cases of acute upper respiratory infection is understandable, it is better to carry out this act of relief before it becomes one of desperate emergency. The indication for tracheotomy arises when the symptoms are increasing and the response to treatment is poor or absent. The patient will thus be saved much suffering and anxiety, and the exhaustion, which may so easily occur in a toxic or shocked patient and which may end in sudden cardiac failure, will be avoided.

Children

In young children, who are especially liable to the oedematous form of acute laryngitis, the symptoms of dyspnoea are aggravated by the restlessness and terror which easily arise from the difficulty in obtaining sufficient oxygen

The use of sera and sulphonamides is equally valuable in children and in adults, but when the necessity for tracheotomy is in doubt, the child should be placed in an oxygen tent. Many instances have occurred in which the need for tracheotomy has been avoided or a catastrophe averted by so increasing the proportion of oxygen in the inspired air. The use of an oxygen mask may take the place of the tent, but is often more frightening to the child. If the condition becomes worse even with oxygen administration, or does not respond after an hour or so, then tracheotomy should be carried out without further delay.

Local treatment

In typhoid fever the appropriate serum should be administered, and in cases of agranulocytic angina pentnucleotides should be given. In addition, local

treatment to the larynx consists in dry inhalations, by means of a suitable respirator, of volatile oils, sprays of an oily solution of Chloretone, and the application of cold. The necessity for tracheotomy must be decided on the individual merits of each case.

(2) Chronic laryngitis

Definition Chronic laryngitis is a persistent low-grade inflammation of the mucous membrane of the larynx, characterized by redness and swelling.

Aetiology This condition may follow one or more attacks of acute laryngitis. It is often associated with a similar condition in the pharynx—chronic pharyngitis—and in many patients it follows a purulent infection of the nose and paranasal sinuses, of the tonsils or of the teeth. It is more common in men over the age of 40, especially when there is over-use of tobacco and alcohol and when there is persistent over-use of the voice as in the case of street hawkers, itinerant showmen and those with similar occupations. Working in any atmosphere laden with irritant fumes, such as in foundries and in works during certain chemical processes, may lead to the condition.

Clinical picture There is persistent hoarseness which increases with use of the voice during the day. Pain is often complained of and is of the congestive type. It is such as to cause dysphagia in a number of cases. There is also a constant hawking and coughing—clearing the throat—in an attempt to remove small traces of adherent mucus.

Diagnosis On examination, the mucous membrane is uniformly thickened and a deeper red than usual. The vestibular folds are swollen and may partially or wholly hide the vocal folds from view. An important diagnostic point is the symmetry of the changes in the larynx; the two sides, and very often all parts of the larynx, being equally affected, in contrast to the asymmetry of the changes in the specific infections and neoplastic changes The condition is commonest in the age-groups in which carcinoma is met, and a thorough examination of every part of the laryngeal mucosa should be carried out before the diagnosis of chronic laryngitis is made.

Treatment.—The treatment consists primarily in removing the causative lesion in the mouth and upper air passages, and in regulating the use of the voice and the misuse of alcohol and tobacco.

The use of gargles and the spraying of astringent fluids by a laryngeal atomizer are of value. The most valuable of the astringents are zinc chloride and iron perchloride in watery solution in a strength of 4 grains to the ounce. Much benefit may be obtained by the application to the interior of the larynx of a 15–25 per cent solution of Argyrol in water by means of a laryngeal syringe. The volatile oils sprayed into the larynx are also useful; Chloretone being probably the best medicament Surgical measures are not required for this condition.

Varieties of chronic laryngitis

(i) *Hypertrophic laryngitis.*—As its name implies, hypertrophic laryngitis is a type of chronic inflammation in which the mucous membrane is thickened. This is particularly noticeable on the vestibular folds, the arytenoid processes and the interarytenoid region. In some cases a mass of thickened mucosa is formed at the posterior ends of the vocal folds causing very severe hoarseness. This condition in its more exaggerated form with much thickening of the

epidermal layer is known as pachydermia of the larynx. Hypertrophic laryngitis does not respond to the methods suggested for chronic laryngitis and it may be necessary to apply the galvano-cautery to the masses of thickened membrane or to remove them by forceps through a direct laryngoscope.

(ii) *Polypoid laryngitis.*—This condition consists of polypoid swellings of the upper surfaces of the vocal folds and along their edges. The projection is somewhat fusiform; it begins at the anterior commissure and spreads posteriorly, often for two-thirds of the length, or it may reach the vocal process The condition is usually symmetrical, but may be restricted to one side. Hoarseness is the invariable and only symptom.

Hoarseness

Treatment consists in eliminating outside causes of inflammation, but removal of the polypoid swelling by forceps under direct endoscopy is usually necessary. The swellings peel off the vocal folds readily but care should be exercised not to injure the vocal fold proper, especially if both sides are being treated at the same time, so as to reduce the risk of adhesion and constriction of the glottis. Ideally the two sides should be treated separately at intervals of from 7 to 10 days.

Terminology

(iii) *Laryngitis sicca* (*atrophic laryngitis*).—This is a dry condition of the laryngeal mucosa with adherent muco-purulent crusts. It almost invariably results from suppuration in the nose and paranasal sinuses, so much so that the term rhinitic laryngitis has been applied to it. It may also be associated with atrophic rhinitis with ozaena.

The mucous membrane of the larynx is rough and congested, particularly in the posterior part of the glottis. The membrane of the interarytenoid space is thrown up into a series of small folds and in this region there is almost always an adherent plaque of inspissated muco-pus which lies on the upper surface and seems to spill over through the posterior commissure into the subglottic region. The crusts are often attached to the ventricular folds and to the vocal folds on one or both sides. Similar adherent patches will be seen on the posterior wall of the pharynx and nasopharynx. Hoarseness is common, and a feeling of constriction or even of something stuck in the throat is a common complaint. Attempts to clear out the muco-pus by coughing and hawking are made and sometimes, as a result, a little blood is brought up.

Treatment

Treatment consists mainly in clearing up the nasal suppuration by whatever means are necessary and the muco-purulent crusts and the inflammation will often disappear. It may be necessary to take more active steps in the larynx to remove the crusting, and for this purpose syringing out the larynx, while the patient phonates, with 2 cubic centimetres of a solution of sodium bicarbonate (40 grains to the ounce) will clear away the crusts. If this fails, removal by forceps through an endoscopic tube is required.

(iv) *Keratosis laryngis.*—This is a condition in which heaped-up masses of a dead-white appearance occur—usually on the edge and upper surface of one or, more rarely, both vocal folds. These masses consist of large numbers of cells from the horny layer of the epithelium. There has been a localized proliferation of the horny layer, without any corresponding proliferation of the cells of the dermal layer.

There are usually several separate areas covered by the white masses which are irregular in shape and distribution. They tend to grow very slowly but may eventually coalesce.

The only symptom is hoarseness, which is not very severe, and there is no limitation of the movement of the vocal fold.

As they tend to occur in late middle-life the masses may be mistaken for an epithelioma but they are of a purer white colour than is a cancer and are seen to be entirely superficial.

It is often suggested that rest to the larynx and avoidance of irritation by tobacco and other means, perhaps with the addition of a spray, as for chronic laryngitis, will cause them to disappear, but once the masses of horny epithelial cells have formed they usually need to be removed; this is carried out by picking off each individual mass with curette-ended forceps through a direct endoscopic tube. Complete removal needs patience and perhaps more than one sitting, but once all the masses are removed recurrence is uncommon. There is no evidence that this is a pre-cancerous condition but observation should be kept up for a year or more after removal. *Removal*

(v) *Contact ulcer.*—A contact ulcer is a shallow ulceration occurring on the medial aspect of one or both vocal processes. It is said to be commonest in those who use their voices continuously over long periods, but the ulcer also occurs in individuals who have never used the voice to excess. The ulcer appears to be due to the pressure exerted by the two vocal processes on one another.

The appearance is that of a cup-shaped depression with a rather thickened rim on one vocal process. The opposite vocal process shows a knob-shaped eminence which fits into the cup, and which may be a purely anatomical structure or may be a small granuloma formed in another contact ulcer. The only symptom is slight hoarseness; rarely there is loss of voice. The ulcers will disappear and heal in the majority of cases if a period of several weeks' silence is observed, and healing may be assisted by the spraying into the larynx of an oily solution of menthol, Chloretone or eucalyptus oil or the syringing into the larynx at daily, or longer, intervals of a 15–25 per cent aqueous solution of Argyrol.

(vi) *Perichondritis.*—This consists of an inflammatory swelling of the perichondrium of one or more of the cartilages of the larynx. It is characterized by the infiltration and swelling of the fibrous part of the perichondrium which causes much swelling in the overlying areolar and submucous layers, and causes the death of the cartilage-forming layer of cells which in turn gives rise to necrosis of the cartilage itself.

Perichondritis may follow injury by direct violence, by penetrating wounds, or as a result of one of the acute exanthems, typhoid fever, diphtheria or influenza. It also occurs not infrequently in tuberculosis or syphilis, and may be caused by the injudicious application of x-rays or radium. Localized perichondritis sometimes occurs after the operation of laryngofissure. *Cause*

The appearance in the larynx is that of considerable oedematous swelling in the region appropriate to the part of the laryngeal skeleton which is involved. The swelling immobilizes one vocal fold and sometimes both, and causes loss of voice rather than hoarseness. Pain, due to congestion and tension, is present and there is always a tendency to dyspnoea. Oedema may cause a dangerous degree of obstruction necessitating tracheotomy. An abscess may form between the perichondrium and the cartilage with some necrosis of the latter, causing even more swelling, more pain and often more dyspnoea. An abscess *Loss of voice*

is commonly found in association with one of the arytenoid cartilages and eventually the abscess ruptures, discharging pus, and in due course extruding the necrotic cartilage. The final result of infection in this region, with or without the death of the cartilage, is fixation of the crico-arytenoid joint and immobility of the vocal fold.

Perichondritis of the wing of the thyroid cartilage may result in a necrotic perforation of the cartilage with abscess formation on the outer aspect of the cartilage. This leads to a painful swelling which is covered by the infra-hyoid muscles.

The swelling superficial to the thyroid cartilage should be incised and drained when the presence of pus is established, but healing will usually be slow on account of the sequestration of portions of cartilage.

The sulphonamides have not had any degree of success in perichondritis, but it is probable that heavy intramuscular doses of penicillin will produce a resolution, at any rate in the early stages.

The end-results of perichondritis are usually hoarseness from fixation, and also, but less commonly, cicatricial stenosis of the larynx. If much cartilage has been lost the stenosis may be prevented or reduced after tracheotomy, by fitting into the lumen of the larynx a plastic moulded splint which must be worn for some weeks until the larynx has assumed a stationary condition.

(3) The infective granulomas

(a) *Tuberculosis*

Definition Tuberculosis of the larynx consists of an infiltration of the submucous tissue by tubercles, and of the various manifestations of the disease which follow.

Classification Tuberculous disease, as it occurs in a mucous membrane, may be divided according to Aschoff into four stages: (1) infiltration, which may be further subdivided as to whether or not there is an associated oedema; (2) ulceration; (3) perichondritis, and (4) tumour formation.

Infiltration without oedema represents the subacute type of the disease in which the nodules are separate and may be distinguished through the surface of the membrane. Infiltration with oedema represents the more active type of the disease. In this type more tubercles are being formed rapidly and are growing. There is more round-celled infiltration immediately surrounding the tubercles, and swelling of the mucous membrane is caused by the formation of lymph spaces—small ones in close proximity to the tubercles and larger ones in the connective tissue at a distance from the tubercles. This condition has been described as a lymphangitis, and when the swelling has reached a sufficient degree the surface becomes smooth and the tubercles are no longer visible.

Oedema Oedema is a sign not only of increased activity of the disease but, especially when it occurs in long-standing cases, may be an indication of reduced resistance.

Ulceration. If the tubercle spreads towards the surface, it erodes the epithelial layer and an area of granulation tissue appears on the surface and forms an ulcer which at first is usually circular, but which becomes irregular in shape owing to eccentric extension or to the coalescence of two or more ulcers. The ulcer is shallow, with a granular floor, rather undermined edges and congestion around its margin. Mucus is almost invariably adherent to the

floor of the ulcer and can readily be wiped away, leaving a granular base which tends to bleed easily. Tuberculous ulcers are always productive of pain. The most sensitive part is the growing edge and particularly its line of junction with the floor of the ulcer. The passage of food, drink or saliva over the surface of an ulcer always gives rise to pain.

Perichondritis is caused by extension of the inflammatory process from the submucous layer to the perichondrium. It tends to occur in the more active types of the disease, especially when there is ulceration and secondary infiltration; this has been considered to be due to deposition of tubercle bacilli by the blood stream at the level of the perichondrium. It is characterized by a considerable small round-celled infiltration with many tuberculous nodules and a great deal of oedema. The perichondrium is stripped off the cartilage, the nutrition of the latter being impaired and necrosis taking place. Necrosis, as in the non-tuberculous perichondritis, usually involves the arytenoid and epiglottic cartilages, with sequestration of the former and either quiet absorption or necrotic separation of the latter.

Clinical appearances

The clinical appearances are those of great oedematous swelling which may effectually hide all the normal landmarks and contours of the larynx The glottis may be reduced so that respiration becomes embarrassed. Perichondritis is the commonest cause of such degree of dyspnoea as to make tracheotomy necessary.

Subsidence of the oedematous swelling in a healing perichondritis usually leaves some permanent degree of thickening and it also results in distortion of the epiglottis and, at least, in partial fixation of the crico-arytenoid joint.

Tumour formation consists of a mass of tuberculous tissue with many giant-cell formations. It usually occurs at the posterior end of the vocal and ventricular folds and also in the interarytenoid region. There are two varieties: the granulotuberculoma, which consists of soft tuberculous granulation tissue which tends to bleed and occurs mostly on the vocal processes, and the fibrotuberculoma, which is more solid, with an epithelial covering which forms irregular masses on the posterior ends of the vocal folds or a rounded or pyramidal mass in the interarytenoid space. Sometimes these masses are so tough and leathery that they have been termed pachydermia, though tuberculous in nature.

Granulo-tuberculoma

Fibro-tuberculoma

Intrinsic muscular weakness In addition to the types of the disease in Aschoff's classification there is a frequent cause of hoarseness in an inflammatory change in the internal thyro-arytenoid muscle. Occasionally, there are areas of tuberculous infiltration in the muscle mass, but often there is only a simple inflammatory change with small round-celled infiltration and fragmentation of the muscle fibres. On examination the cords are seen to be bowed on phonation and do not come into apposition except at the two extremities.

Hoarseness

(i) *Symptoms.*—The commonest symptom is hoarseness Any change taking place in the vocal fold prevents its proper vibration and perhaps also the apposition of the two folds. Lesions of the vestibular folds, of the subglottic region, of the arytenoid processes or of the interarytenoid region have the same effect. Lesions restricted to the epiglottis do not cause hoarseness.

Pain

Pain comes second in the order of frequency and may consist of an ache which is due to congestion and is worse after using the voice. A more severe and more serious type is that caused by ulceration. When the ulcer is on the

vocal or vestibular folds it is not exposed to irritation by the passage of food, drink or saliva. Pain may be caused during deglutition because the ulcer is compressed by the sphincter action of the larynx. The pain may be referred to the ear by a reflex along the auricular branch of the vagus nerve. When the ulcer is on the epiglottis, aryepiglottic fold or arytenoid process it is irritated by everything that is swallowed and severe pain is caused. Pain may be so severe that swallowing becomes almost impossible and the patient's resistance is likely to suffer from lack of nutrition.

Dyspnoea

The third symptom, dyspnoea, is the least common but is the most dangerous. It is caused by oedematous swelling of the vestibular folds, the arytenoids, and sometimes of the epiglottis. It may be caused by masses of fibrotuberculoma in the posterior part of the glottis, by paralysis of both recurrent laryngeal nerves or by fixation of both crico-arytenoid joints. It rarely becomes marked enough to necessitate tracheotomy, but is usually an indication that the disease has passed the stage of probable recovery.

General

(ii) *Prognosis.*—The expectation of life of a patient suffering from tuberculous laryngitis is less than that of a patient suffering from a comparable degree of disease of the lungs but with a healthy larynx. In young adults and adolescents resistance is at its lowest, and the prognosis is very bad in these age-groups. In patients over 40 years of age the resistance is greater and the disease takes on a more quiescent form, but may suddenly give way with very rapid deterioration.

Local

The prospect of complete recovery in the larynx depends upon the resistance and the progress of the disease of the lungs, but it also depends upon the type and degree of the lesions in the larynx. Infiltration with or without oedema may disappear entirely and leave a normal voice. Ulceration may heal and leave a small scar but unimpaired function. Massive swelling of the ventricular folds rarely disappears completely and some hoarseness persists. Perichondritis leaves some swelling and often deformity. Unless restricted to the epiglottis this will result in persistence of some degree of hoarseness.

(iii) *Treatment.*—The treatment of tuberculosis of the larynx consists in general and local measures. The methods of immobilizing the lung and the vocal fold and applying medicaments to the larynx are described in detail in the appropriate works of reference. Special mention may be made of the methods used to combat pain, on swallowing, due to ulceration of the laryngeal mucosa. Aspiration through a suitably bent glass tube (Leduc) of ½ a drachm of equal parts of benzocaine and orthocaine 15 minutes before taking food gives considerable relief. As an alternative an emulsion of benzocaine is almost equally helpful in cases in which the powder is irritating and causes coughing.

Galvano-cautery

The galvano-cautery is an extremely useful method of treating ulceration, infiltration and non-oedematous swelling in the larynx, and is one of the most certain measures for alleviating pain. It is carried out after cocainization by the indirect laryngoscopic method, or may be performed through the direct laryngoscopic tube, though this should be unnecessary. The platinum wire, heated to a bright red, is plunged into the nodule or into the thickened arytenoid process, ventricular fold or other part and after being held for 2 or 3 seconds is withdrawn still heated, and if necessary plunged in again in another place. If the wire is allowed to cool before withdrawal it becomes adherent

and some laceration and bleeding will be caused before it is freed. For the relief of pain the cautery is applied to the undermined edges of the ulcerated areas—the edge being the most sensitive part of the ulcer.

When pain is not relieved by analgesic applications or by galvano-cautery, the method of injecting the superior laryngeal nerve with alcohol will often produce complete relief. This injection gives anaesthesia which will last from 3 weeks to 2 months, after which it can be repeated if necessary. It is usually done on one side, but both nerves may be injected at the same time when both sides are badly involved. The nerve is found immediately above the postero-superior end of the oblique ridge on the side of the thyroid ala.

Alcohol injection

The injection is made with a 2 cubic centimetre syringe and a needle about 4 centimetres long which has a groove scored on it 1 centimetre from the point. One cubic centimetre of a solution containing 2 per cent of eucaine in 80 per cent alcohol is drawn up into the syringe. The oblique ridge is identified with the thumb nail and the needle is entered at this point at right angles to the skin and pushed in to a depth of 1 centimetre. The point of the needle is gently moved in and out until the patient experiences a pain which is referred to the ear. This indicates that the nerve has been entered and the referred pain along Arnold's nerve elicited. The piston is withdrawn a little to make certain that the needle is not in the sinus piriformis, when air would enter the syringe, or in a blood-vessel, when blood would appear in the barrel The patient has been previously instructed to raise the hand when he feels the pain and to lower it when the pain passes off. The contents of the syringe—1 cubic centimetre of the solution of eucaine in alcohol—are now injected and there is usually a recurrence of pain. This passes off very quickly and the needle can be withdrawn.

It is usual to apply a small collodion dressing to the puncture.

During the whole of this procedure the patient must keep very quiet and must neither speak nor swallow as these actions move the larynx upwards and the anatomical relationships are lost. The syringe and needle, once the nerve has been entered, must also be kept very steady and not moved in any way.

The effect of this injection is instantaneous and it is usual to give a drink to the patient immediately the dressing has been applied. The patient finds that he can swallow quite comfortably and his first reaction expresses itself in a look of astonishment and gratitude.

Alternatively, the nerve may be exposed through an incision which runs forward from the anterior margin of the sternomastoid muscle at the level of the greater cornu of the hyoid and is carried forward in the direction of the thyroid notch. The dissection is deepened until the posterior margin of the thyrohyoid muscle is found. The nerve is found lying immediately superior to the superior laryngeal artery and passes forward under the muscle. The nerve may be injected with alcohol under vision, but in cases in which alcohol injection has not produced the desired results the nerve may be divided between ligatures and a portion excised The result is complete and permanent anaesthesia of the appropriate area of the interior of the larynx. The only bad result from the anaesthesia is that the larynx is a little more liable to inflammatory processes and to the spread of existing ones, but the cough reflexes are not abolished.

Excision of nerve

The method may fail in those patients whose ulceration lies in, or spreads

into, the area innervated by the glossopharyngeal nerve—the outer aspects of the arytenoid and aryepiglottic region and the free portion of the epiglottis.

(*b*) *Syphilis*

Definition

Syphilis of the larynx is the laryngeal manifestation of a *general systemic* syphilitic infection.

Primary infection is almost unknown and secondary manifestations are less *commonly met than tertiary lesions.*

Aetiology

Laryngeal syphilis is more common in acquired than in congenital syphilis. The congenital type may give rise to lesions in the first few months of life. These are of a secondary nature and may be followed by tertiary lesions. In congenital syphilis the tertiary lesion may occur in adult life without any previously recognized secondary lesion. *In acquired syphilis the secondary* lesion may occur before the extralaryngeal primary sore has healed, or its onset may be delayed some 6 or 8 weeks. The tertiary lesion may occur at any period, but usually does not appear during the first 2 or 3 years after infection.

Secondary infection

Secondary infection, which is probably common in a transitory phase with hoarseness for a few days while the skin eruption is present, is not often sustained enough to call for the attention of the laryngologist. It assumes the *form of a hyperaemia and thickening of the mucous membrane, with sodden* or oedematous areas known as mucous patches. The only symptom is hoarseness. The condition often disappears without treatment, but under specific treatment it clears up very rapidly and leaves no trace behind.

Tertiary syphilis

Tertiary syphilis may take on one of a number of forms. The commonest is the ulcer which occurs on the epiglottis or on the posterior end of the ventricular fold and the contiguous arytenoid process. It is usually single and characterized by its raised hyperaemic margin, its sharp-cut edge and its *sloughing base. Though not often circular its edges show a smooth regular* outline. The ulcer extends deeply and may reach underlying perichondrium but it does not extend very much in a peripheral direction and, therefore, destruction is not widespread. There is usually some *destruction of cartilage* by necrosis when the ulcers form on the epiglottis.

Hypertrophic type

The hypertrophic type takes the form of fibrogranulomatous masses which form on the vocal folds, in the laryngeal sinus and on the vestibular fold. It is often bilateral, fairly symmetrical and tends to preponderate in the posterior *half of the glottis.* The masses are firm and suggestive of pachydermia, but are more extensive. They cause hoarseness of a rough, harsh type, and not infrequently give rise to such obstruction of the glottis as to require tracheotomy. They are sometimes *associated with gummatous deposits in the lung.*

Perichondritis

Perichondritis may be due to the extension of a ummatous ulcer, or to the isolated involvement of the perichondrium of o r cartilage—usually the epiglottis or arytenoid. *It is characterized ematous swelling,* and is followed ge and deformity o is, by ankylosis of the crico-aryte d by the sequestra ytenoid cartilage.

Treatment

Treatment co 'application of vi philitic re dies with spraying o larynx in ulcerat derable ment *occurs* ve complete reso There s some los h scarring and ontra

following the fibrogranuloma, persistence of hypertrophic swelling. Dyspnoea may be caused by the great swelling in perichondritis, or as a result of the encroachment on the glottis of the fibrogranulomatous hypertrophy. Tracheotomy is necessary in many cases, especially in the latter type. When there is any swelling in the larynx the administration of iodides should be carried out with the greatest care, as a rapid increase of the swelling may take place. Iodides are better omitted in syphilis of the larynx, but if used small doses should be given at first, and gradually increased, the larynx being watched throughout treatment. When tracheotomy has already been performed iodides may be given with greater freedom.

(c) Scleroma

Scleroma, or rhinoscleroma, is a diffuse nodular hypertrophy of the mucous membrane of the nose, pharynx, larynx, trachea and bronchi. It was originally peculiar to the Balkan countries but spread to Poland and Hungary and, because of the dispersal of considerable numbers of people from eastern Europe, is appearing in many parts of the world

Clinical signs In the larynx the appearance is that of pale rounded swellings which are fairly symmetrical and which occur mainly in the subglottic region, but spread to the vocal and vestibular folds. The lesions are characterized by a firm and resistant consistency. *Symptoms* Hoarseness is invariable and severe; dyspnoea occurs later and increases slowly. The disease may be present for 10 years or more without inconveniencing the patient.

Treatment The condition is very resistant to all forms of treatment. The most hopeful is irradiation either by a high-voltage x-ray apparatus or by radium. Frequent repetition of a number of small doses of x-rays or the application of radium to the side of the larynx has given relief.

When isolated areas of swollen tissue are obstructing the airway they may be removed with very little bleeding. These localized masses tend to recur but only very slowly, and considerable relief will be obtained for long periods.

(d) Leprosy

Leprosy of the larynx may occur in an individual who has suffered for some years from leprosy elsewhere in the body and whose nose and pharynx are already involved.

The disease in the larynx may be of the atrophic or anaesthetic type, or of the nodular type, and most commonly involves the epiglottis and the anterior commissure.

Prognosis The disease is progressive, but the general condition is likely to cause death before the laryngeal disease has reached a dangerous stage.

Treatment An alkaline spray or lavage with a laryngeal syringe may help to keep the airway clear of crusts and secretion—which, it should be remembered, are highly infectious. Very rarely tracheotomy may be required.

(e) Actinomycosis

Actinomycosis is a very rare condition in the larynx and is almost always a direct extension of the condition in the mouth or jaws. It is manifested by considerable swelling which may embarrass respiration. Oedema of the

larynx may occur in the presence of an infection in the floor of the mouth or neck and will lower the prospects of recovery.

4. NEOPLASTIC DISEASES

(1) Benign tumours

Neoplasms of benign nature occur in the interior of the larynx with about the same frequency as intrinsic malignant tumours, but much less frequently than all the types of malignant growth of the larynx (Plate III). They occur more often in men than in women, and the majority are found in young and middle adult life.

Age and sex incidence

(a) *Single papillomas*

The single papilloma occurs on the vocal fold usually about the junction of the anterior and middle thirds and takes the form of a soft pedunculated tumour arising from the edge of the fold. It may assume a somewhat sausage-like shape, or it may divide into two or three projections like the fingers of a glove. The tumour is usually so soft that it hangs down into the subglottic space during ordinary quiet respiration but is carried up above the level of the folds during the forced expiration of phonating and coughing. It is attached superficially and there is no infiltration of the vocal fold.

Histology

Histologically the single papilloma consists of a central dermal skeleton covered by epidermal cells many layers thick.

It causes a rough hoarseness due to its being set into vibration by the expiratory current or to its being impacted between the vocal folds. The voice changes much because of the assumption of various positions of the tumour in the larynx, and may occasionally be normal for a few syllables owing to the tumour remaining in the subglottic space. It causes no other symptoms.

Treatment.—The treatment consists in removal, and this may be carried out by indirect laryngoscopy under local cocaine anaesthesia, using a laryngeal mirror and curved laryngeal forceps, or by using the direct endoscopic tube and straight laryngeal forceps under local or general anaesthesia. Curette-ended forceps should be used and pressure of the two cutting edges is usually sufficient to separate the tumour from the vocal fold. If not, a slight avulsive twist will suffice to free it. There is a minimal amount of bleeding. There is little tendency to recurrence, and if the tumour does return it is usually in the presence of nasal, oral or pharyngeal sepsis.

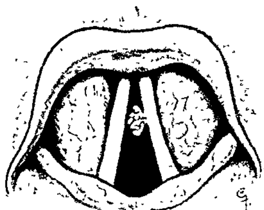
Freeing of tumour

(b) *Multiple papillomas*

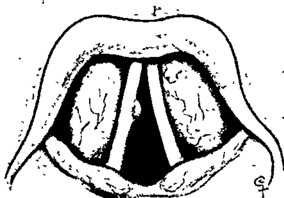
These occur in young children, usually appearing in the first 5 years of life. In structure they resemble the single papilloma but in appearance they are rather flatter and compare with the harder warty papilloma of the skin. They occur in considerable numbers on the vocal and vestibular folds, may be found inside the laryngeal sinus and may spread into the subglottic region and trachea, and on to the arytenoid processes and aryepiglottic folds. A few may reach the epiglottis and even involve the pharyngeal mucosa. The mass of these papillomas is so considerable that not only is hoarseness very marked, but dyspnoea is frequent, and the majority of patients require tracheotomy at least once during the course of the disease.

Dyspnoea frequent

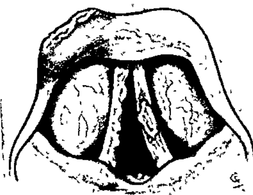
The papillomas may be removed by a number of methods but recurrence is almost invariable. Removal of the growth and other forms of treatment may



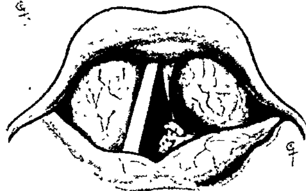
(a)



(b)



(c)



(d)

PLATE III

(a) Papilloma of vocal fold. (b) Soft fibroma of vocal fold. (c) Composite drawing of tuberculous laryngitis. (d) Tuberculous laryngitis

be repeated, and if the patient survives the recurrent emergencies and intercurrent disease, the papillomas tend to cease forming during the period of adolescence.

Some patients are first seen on account of hoarseness; many are brought for advice only when dyspnoea is becoming urgent. Examination will show the glottis almost filled with papillomatous masses and often the airway is not visible. In either case a tracheotomy should be performed as a first measure, and should be done as far away from the larynx as possible to prevent the tracheal opening becoming blocked by an extension of the papillomas. Division of the thyroid isthmus will give the necessary distance. Having made respiration safe, the papillomas may then be removed by means of forceps through a direct endoscopic tube, or by suspension laryngoscopy. The removal must be carried out carefully and methodically—clearing each separate part of the larynx of all tumours. It will often be worth while doing this on more than one occasion, partly because after a time blood begins to obscure the field, and it is rarely possible to remove all the papillomas at one sitting. Care must be taken not to injure the mucous covering of the larynx, particularly on the vocal and ventricular folds. Injury will reduce the probability of a good voice at a later stage and also causes the risk of adhesion between the two sides and the formation of webs. It is doubtful whether the application of acids and caustics prevents recurrence; their use is more certain to damage the mucous membrane and cause scarring.

Tracheotomy

In a number of patients there will be so many polypi below the vocal cords that removal is impossible except by performing a thyrotomy or laryngofissure—the details of which are described later (*see* page 361). The papillomas can be very thoroughly removed by this means, but it is a much more serious operation than endoscopic removal and should be reserved for cases with subglottic extension or adherent vocal and vestibular folds.

Prognosis.—The prognosis of multiple papillomas is not good. The repeated operations on the larynx and the necessity of opening the trachea on a number of occasions, although not immediately fatal, reduce the child's resistance to infection, and death often occurs from broncho-pneumonia or other intercurrent malady. Survival into the "teens" holds the prospect of ultimate recovery with, however, a persistent hoarseness and probably some stenosis in the larynx.

(c) *Fibromas*

Under this heading may be included the true fibroma, the so-called soft fibroma and the singer's nodes.

The true fibroma is an uncommon tumour consisting of a mass of fibrous tissue which becomes pedunculated as it increases in size and becomes ovoid or spherical with a diameter up to ½ inch. It frequently arises at the anterior commissure, hangs down into the trachea and is blown up into the vestibule of the larynx during expiration and phonation. It causes hoarseness and may embarrass breathing.

It is very easily removed by forceps through an endoscopic tube or by indirect laryngoscopy and, as a rule, does not recur.

The soft fibroma (so-called) is a sessile hemispheroidal swelling occurring on the edge of the vocal fold, usually in the middle third. After the single

Soft fibroma

papilloma, it is the commonest benign tumour in the larynx—if it can truly be called a tumour—and occurs in both sexes and at any adult age. It causes hoarseness, preventing proper apposition of the vocal folds. It is recognized by its smooth surface and by its sessile hemispheroidal appearance. It can be removed readily by indirect or direct endoscopy, but as it is often difficult to engage by curette-ended forceps, punch-ended forceps should be used. Recovery of appearance and function of the fold is perfect and recurrence is rare.

Structure

The histological structure of this tumour is a central cystic space containing some degenerate red blood corpuscles, surrounded by a layer of fibrous tissue and a layer of keratinized cells, and finally covered by the stratified epithelium of the fold. This structure suggests that its origin is a small haemorrhage under the mucous membrane which has not been completely absorbed and, acting as a foreign body, causes—by irritation—the formation of a thick-walled cyst.

The singer's nodes are symmetrical nodules occurring on the free edge of the vocal fold and causing hoarseness. They occur in the middle third and usually posterior to the midpoint, are often only the size of a pin's head and rarely as much as $\frac{1}{8}$ inch in diameter. They occur in singers, lecturers, public speakers, and those who use the voice wrongly or excessively. Faulty voice production is probably the commonest cause. They are white, hemispherical and sessile and on examination consist of hypertrophied epidermal elements with some fibrous tissue cells.

Size

If only very small they may be treated by prolonged rest and correction of voice production faults, but if this fails after 4 or more weeks, or if, as is often the case, quick results are desired, the nodules can be removed by curette-ended forceps—preferably through the endoscopic tube, great care being taken not to injure the vocal fold itself. Very small nodules can be destroyed by the galvano-cautery.

(*d*) *Cysts*

Cysts may be congenital, or may be formed by retention of secretion in mucous glands

Congenital cysts are usually associated with the laryngeal sinus or the saccule. They may distend the vestibular fold until the entire lumen of the larynx is occupied or they may appear on the internal aspect of the arytenoid process or aryepiglottic fold. Sometimes the swelling may have an intralaryngeal portion and a second portion in the medial wall of the piriform fossa, or even on the outer aspect of the thyroid cartilage. The cyst may lie dormant for years and then increase in size very rapidly. Puncture and aspiration of the fluid, or excision of considerable amounts of the wall, is followed by very rapid recurrence of swelling. Excision is necessary and is best performed by a transthyroid pharyngotomy (*see* page 369) and deliberate dissection by this approach. When there is an aperture through the thyroid cartilage the extra-laryngeal portion of the cyst will also require excision.

Puncture and aspiration

Retention cysts occur most commonly in the valleculae where they may attain such a size as to overhang the epiglottis and to be visible over the base of the tongue, and on the epiglottis where they are usually small and perhaps multiple.

The small cysts on the epiglottis rarely cause symptoms and do not need treatment.

The vallecular cysts cause discomfort and a feeling of a large swelling in the throat. There may be some slight degree of interference with deglutition but never to any serious extent.

This type of cyst is caused by the accumulation of mucoid secretion, and the gradual distention of the gland. Puncture and aspiration are only followed by refilling. Removal of as much of the wall as possible by means of punch forceps followed by scraping of the remainder by dry gauze swabs will often effect a cure. This may be done through the mouth by using an endoscopic tube, or a Davis's gag and tongue depressor as for dissection of tonsils.

(e) *Angiomas*

Benign neoplasms consisting of vascular tissue are uncommon but may occur in any part of the larynx. They are almost all haemangiomas and may vary from large ones occupying almost half the larynx, and consisting of large cavernous blood spaces, to small nodules on the vocal folds and the epiglottis which consist of a small knot of blood-vessels. The symptoms they cause vary with their position and with their size. A small one on the vocal fold causes hoarseness, small ones elsewhere do not cause any symptoms. Wherever arising, the larger ones, if they influence the movement of the vocal folds, will cause hoarseness, otherwise they might interfere with deglutition or respiration, but this is rare. If they become infected and ulcerated, haemorrhage may occur but this is also rare. The majority do not cause symptoms.

Symptoms

The appearance of these tumours is of a dark-red swelling of irregular surface and irregular shape which gives the impression of being half buried in the tissue in which it is situated, the other half projecting from the surface. On the vocal folds the small haemangiomas are almost pedunculated.

Appearance

Removal of the small ones can be carried out by curette-ended forceps by either indirect or direct laryngoscopy, but preparation should always be made to deal with a persistent haemorrhage. If this is slight it can probably be controlled by application of the galvano-cautery; if more free it may be necessary to perform a tracheotomy and to pack the larynx with gauze. Alternatively small haemangiomas may be destroyed by the galvano-cautery or by diathermy. The large ones are best left alone. They cause mild symptoms and grow very slowly, if at all. Removal, if complete, would destroy the usefulness of the larynx, by disappearance of part of its tissue, and possibly also by scarring and distortion. The great danger would be of haemorrhage which in the case of these large tumours might be cataclysmal and uncontrollable.

Removal

Haemorrhage

(f) *Chondromas*

The tumours consisting of cartilage cells arise almost always in the subglottic region, from the inner face of the thyroid ala or from the cricoid cartilage. They take the form of a smooth, round, hard mass gradually extending across the lumen of the subglottic space, causing hoarseness at an early stage and dyspnoea later.

The only treatment consists in surgical removal and this must be done by external operation. A preliminary tracheotomy should be performed, if not already in existence. Thyrotomy or laryngofissure is carried out (*see* page 361) and the tumour exposed. It may be necessary to divide the cricoid ring to

Treatment

gain a complete approach. The tumours can usually be shelled out by using an appropriately-shaped periosteal elevator and, if necessary, a little excavation with a gouge at the site of attachment. There is little bleeding and the restoration of the airway and the function is usually perfect.

(g) *Lipomas*

The fatty benign tumours are rare but occur as pedunculated masses of somewhat ovoid shape. They arise, as a rule, from the outer aspect of the aryepiglottic fold, and hang down into the piriform fossa and even into the oesophagus. They rarely cause symptoms but if the pedicle becomes sufficiently elongated, may be brought up into the mouth or drawn into the larynx, causing dyspnoeic attacks. They are easily removed by cutting through the pedicle with a snare or a punch forceps—preferably by direct endoscopy.

(2) *Malignant disease of the larynx*

Definition

Malignant tumours arising in the larynx are almost invariably primary. In the great majority of cases they are carcinomas, sarcomas and endotheliomas being extremely rare. The carcinomas are nearly always squamous-celled epitheliomas, but a few are basal-celled.

They are divided into two main groups: (a) intrinsic tumours, which arise in the interior of the vestibule of the larynx—from the vocal folds, the vestibular folds, the laryngeal sinus and the anterior and posterior commissures—and (b) *extrinsic tumours, which may be divided into the true extrinsic tumours arising on the aryepiglottic fold and the anterior wall of the* post-cricoid region; the hypopharyngeal, including those arising on the lateral wall of the piriform fossa and on the posterior wall of the post-cricoid region; and the epilaryngeal, including the tumours of the epiglottis and vallecula.

Aetiology

Malignant disease in the larynx is a disease of late middle life, being rare *below the age of 40 and commonest between 50 and 60 years of age. It is* commoner in men than in women in the proportion of 8 or 9 to 1, except in the case of the post-cricoid carcinoma, when the proportion is reversed.

The disease occurs in its extrinsic form about one and a half times as frequently as in its intrinsic form.

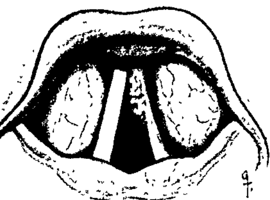
The Plummer-Vinson syndrome *in women may be a precursor of pharyngeal* carcinoma, but no precancerous condition in the larynx has been demonstrated.

(a) *Intrinsic carcinomas*

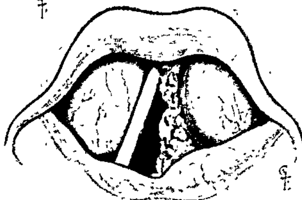
Surgical anatomy

With few *exceptions the intrinsic carcinomas arise in the anterior* and middle thirds of the edge of the vocal fold, the centre of the tumour being always anterior to the midpoint of the fold. This region lies just between the superior and inferior lymph fields of the larynx and is very sparsely supplied with lymphatic vessels and so metastatic dissemination is rare.

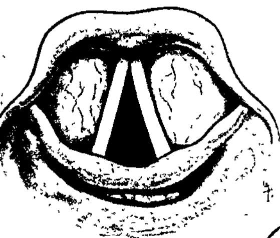
The intrinsic tumour tends *to spread along the edge of the vocal* fold until it reaches the anterior commissure or the arytenoid eminence. It may easily cross over the midline anteriorly and spread on to the opposite cord. Posteriorly it does not often cross over to the opposite side, but rather tends to spread up onto the vestibular fold and the aryepiglottic fold. Anteriorly the tumour extends downwards *into the subglottic region and only* occasionally enters the laryngeal sinus or reaches the vestibular fold. (Plate IV)



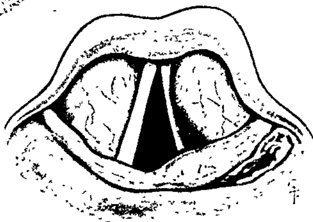
(a)



(b)



(c)



(d)

PLATE IV

(a) Early carcinoma of vocal fold. (b) Advanced carcinoma of vocal fold. (c) Post-cricoid carcinoma. (d) Carcinoma of piriform fossa.

(*b*) *Extrinsic carcinomas*

The extrinsic carcinoma when occurring on the epiglottis extends very slowly but forms a fungating mass. The epiglottis is not well supplied with lymphatic vessels, and secondary deposits do not occur readily. Tumours occurring in the vallecula tend to spread slowly on to the epiglottis but infiltrate deep into the mass of the posterior part of the tongue. Secondary deposits occur in the upper deep cervical lymphatic glands.

The tumours arising on the outer aspect of the aryepiglottic fold tend to spread round onto the lateral wall of the piriform fossa, to surmount the aryepiglottic fold and enter the interior of the larynx. Those arising on the lateral wall of the piriform fossa spread upwards onto the lateral wall of the pharynx and also across onto the aryepiglottic fold Lymphatic spread is free and leads to the upper deep cervical glands.

The post-cricoid tumour arises just inside the mouth of the oesophagus; it may begin on the anterior or on the posterior wall and in either case tends to spread slowly round the circumference. It also spreads upwards onto the arytenoid mass or the posterior pharyngeal wall. The lymphatic drainage leads to the lower deep cervical glands.

Pathology

The tumours occurring in the larynx are almost exclusively squamous-celled epitheliomas and if examined from the point of view of differentiation of cells and frequency of mitotic figures (Broders) it will be found that few of the tumours fall into the fourth group of 75–100 per cent undifferentiated cells, and that the first, second and third groups represent almost equal proportions, the second group perhaps including the greatest number of tumours.

The tumours arising on the vocal fold and on the epiglottis tend to show the highest degree of differentiation—partly explaining their slow progress and late dissemination, the sparse lymphatic drainage being another factor.

The progress of the tumour beneath the surface may be a little in advance of that on the surface, but its deep penetration is slow. A tumour may extend the entire length of the vocal fold before the muscle underlying the vocal fold is infiltrated When the muscle—the internal thyro-arytenoid—becomes involved, the spread of the carcinoma through its whole mass is rapid and the mobility of the vocal fold is brought to an end.

The tumour ulcerates at an early stage and forms a typical saucer-shaped ulcer with nodular everted edges. In the intrinsic growths the ulcer deepens and extends slowly and is only invaded by secondary pyogenic organisms, which produce oedema, at a late stage. In the extrinsic growths the ulceration extends and deepens more rapidly and is infected, with occurrence of oedema, at an early stage.

Metastatic deposits in the upper deep cervical group of lymph glands occur very early, except in the case of the post-cricoid carcinoma.

Symptoms

The first symptom of the intrinsic growth is hoarseness and for a long time there are no other symptoms. It is only in the latest stages that there is any dyspnoea. This may be due to increase in the mass of the growth or to oedema due to secondary infection Haemorrhage may occur in a deeply ulcerated mass but usually is not severe.

In the case of the extrinsic growth the onset of symptoms varies considerably according to the site of origin. Growths on the epiglottis are likely to produce dysphagia and, perhaps, when ulcerated, a little pain. In the later

stages, when they fungate, there will be bleeding and, when the mass becomes large enough, dyspnoea.

The post-cricoid growths give rise to a vague sense of discomfort for some time before real dysphagia sets in—whilst the growth has only spread round a small proportion of the circumference. Later, dysphagia becomes a prominent symptom and is steadily progressive.

Growths on the aryepiglottic and lateral walls of the piriform fossa give rise to dysphagia, hoarseness and a hard mass of metastatic deposits in the neck. The order in which these occur varies—any one of them may present itself first; dysphagia is not commoner than the other two. The hoarseness is due to spread of oedema associated with the very constant secondary infection, which occurs as soon as ulceration takes place.

In many cases inspection by the laryngeal mirror will reveal the nature and site of the lesion. In the intrinsic growth the nodular upper margin of the tumour can always be seen in the anterior part of the fold at the level of its upper surface and the depression of the ulcer is often visible. The extension of the growth at this level can be noted by the mirror, but it is often difficult to see how far it extends into the subglottic space. This can be done by the use of an anterior-commissure laryngoscope, when, by tilting the tube and pressing the healthy fold aside, a view of the subglottic region on the affected side is obtained.

A growth on the epiglottis is immediately seen in the laryngeal mirror and may even be seen by the use of the tongue depressor. With a large epiglottic tumour it may be difficult to see into the interior of the larynx, except by using the direct endoscope.

In growths of the posterior region there is often nothing to see by indirect laryngoscopy, except perhaps a little oedema at the mouth of the oesophagus and some retained secretion in the depth of the hypopharynx. The tumour itself can readily be seen by gently lifting the mass of the larynx away from the posterior wall with the end of a laryngoscope or a short oesophagoscope. The upper margin of the flat ulcerated area can be seen on either the posterior or the anterior wall of the oesophagus. In advanced cases this ulcerated granular mass projects above the level of the post-cricoid sphincter.

Tumours of the aryepiglottic fold and of the piriform fossa may be detected by the mirror in their earliest stages by asymmetry in the sizes of the two piriform fossae. The affected one is shallower and narrower than the other, may contain a little pool of secretion, and there may be oedema of one or other of its walls. The tumour—red granular tissue with an ulcerated area deeper—may be seen lower in the fossa. This may be more completely examined by the direct endoscope.

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Assistance in diagnosing the condition may be obtained by a series of tomographs which will show a mass in one subglottic region, an obliterated laryngeal sinus or ventricle, or a narrowing of a piriform fossa, indicating the pressure of a tumour mass. Otherwise, x-ray films—even by tomography—do [illegible] tumours [illegible] tion [illegible] igns are the interruption in the flow of the bolus at the upper level, and the degree of displacement and distortion of the lumen occasioned by the mass.

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Special aid to diagnosis

Assistance in diagnosing the condition may be obtained by a series of tomographs which will show a mass in one subglottic region, an obliterated laryngeal sinus or ventricle, or a narrowing of a piriform fossa, indicating the pressure of a tumour mass. Otherwise, x-ray films—even by tomography—do not show more than can be seen by laryngoscopy. In the presence of tumours in the piriform fossa and post-cricoid regions, fluoroscopy with the addition of barium boluses will indicate the position and extent of the mass The signs are the interruption in the flow of the bolus at the upper level, and the degree of displacement and distortion of the lumen occasioned by the mass.

tuberculosis or other pulmonary disease—and to examine the sputum for tubercle bacilli. The Wassermann reaction should be investigated, since tertiary syphilis and carcinoma may be very similar in appearance and it is also possible for the two diseases to coexist.

Finally, it is essential that a portion of tumour should be removed for histological examination, partly to confirm the diagnosis however obvious it may appear to the naked eye, and also to attempt to form some estimate, according to the degree of differentiation of cells, of the probable reaction to treatment.

Differential diagnosis

The differential diagnosis between cancer and other lesions of the larynx is contained in the description of the symptoms and clinical appearances of each condition.

For easy recapitulation and to avoid repetition the various points on which diagnosis may be based are set out in the following Table.

TABLE
DIAGNOSTIC CRITERIA

	INTRINSIC CARCINOMA	EXTRINSIC CARCINOMA	TUBERCULOSIS	SYPHILIS	CHRONIC INFLAMMATION
Age	45–65	45–65	20–40	30–50	40–60
Site	Anterior half of vocal fold	Outside larynx	Posterior half of glottis	Epiglottis and posterior half of glottis	Generalized
Surface	Granular	Granular	Smooth, with or without nodules	Smooth or granular	Smooth
Oedema	Late	Early	In acute stage	Marked	Unusual
Ulcer, edge	Everted	Everted	Undermined	Sharp cut	Absent
Ulcer, base	Granular	Granular	Smooth: adherent mucus	Sloughing	Absent
Ulcer, surrounding mucosa	Normal	Normal, oedematous later	Bluish, congested	Reddish	Absent
Pain	Absent	Slight	Marked	Absent	Slight
Dysphagia	Absent	Marked	Frequent	Rare	Rare
Hoarseness	Invariable	Frequent	Almost invariable	Frequent	Frequent
Bleeding	Late	Late	Rare	Absent	Absent
Salivation	Absent	Frequent	In advanced cases excessive	Moderate	Moderate
Secondary deposits in neck	Absent	Early	Absent	Frequent	Absent

Prognosis

The prognosis of intrinsic carcinoma of the larynx is good, better than in many other parts of the body. That of extrinsic carcinoma is extremely bad.

In 70 per cent or more of cases an early tumour of the vocal fold can be cured by operation, by intrinsic irradiation or by distant irradiation. A more

advanced carcinoma, which has not extended outside the anatomical limits of the larynx, may be cured by total laryngectomy in a very high proportion of cases. The extreme rareness of lymphatic metastasis in intralaryngeal growths adds considerably to the expectation of recovery.

(c) *Treatment*

The treatment of carcinoma of the larynx may be surgical removal of the actual tumour and surrounding parts, or it may consist in the application of the *gamma* rays of radium or of high-voltage x-rays to the tumour itself and to any metastatic deposits.

Choice of treatment

The approach and the technique differ according to whether this growth is (a) intrinsic, (b) true extrinsic—aryepiglottic fold or anterior wall of post-cricoid region, (c) hypopharyngeal—outer wall of piriform fossa or posterior wall of post-cricoid region and (d) epilaryngeal or epiglottic.

Intrinsic tumours

(i) *Intrinsic growths.*—When the carcinoma is restricted to one vocal fold and has not reached the arytenoid process, or has only just spread across the anterior commissure to the opposite fold and when the mobility of the fold is unimpaired, the treatment may be of a relatively conservative nature. If, however, the tumour has reached the arytenoid process or has spread over more than half of the opposite vocal fold, or has its main mass in the subglottic region, or has caused immobility of the vocal fold, then the treatment of choice is a total laryngectomy. The conservative measures consist in the local removal through an incision which divides the anterior wall of the larynx and is known as laryngofissure or thyrotomy, the interstitial implantation of radium in needles immediately external to the soft tissues of the larynx, or distant irradiation by teleradium or a deep x-ray apparatus. The decision concerning which of these three measures is adopted is not easy and depends upon the experience of the surgeon. Laryngofissure is probably the most certain, and can be used for rather more advanced cases than interstitial irradiation. It does, however, result in a permanent impairment of the voice which is always rough and husky.

Conservative treatment

Irradiation

Distant irradiation when successful leaves a perfect voice and a normal larynx, but the percentage of cures is not so high; the interstitial method of irradiation has a high percentage of cures—comparable to those of laryngofissure.

When in doubt which of these measures to adopt, it will be wise to perform laryngofissure. If after the fullest examination there is doubt whether or not laryngofissure is practicable, it is wise to perform laryngectomy.

(ii) *True extrinsic growths.*—These are extremely difficult to treat surgically, and respond badly to distant irradiation; this is especially so in the post-cricoid type of growth.

Irradiation

Irradiation causes a temporary recession of the post-cricoid tumour, whether on the anterior or posterior wall, but recurrence is invariable and each recurrence responds less well to further irradiation. The aryepiglottic tumours give a somewhat better response to irradiation and a certain number of cures lasting for 8–10 years have resulted, but in the majority of cases, though they improve for a time, the ulcer and the mass of tumour never quite disappear and speedily recur.

Surgical removal

Surgical removal entails sacrifice of the larynx in either case. For the

postcricoid tumour a portion of the anterior wall of the upper part of the oesophagus may have to be removed and a lengthy process of repair carried out by turning in flaps of cervical skin and creating a new pharyngeal tube. In the case of aryepiglottic tumours laryngectomy may be sufficient but if not, more of the wall of the piriform fossa may have to be removed. This will make closure of the pharynx more difficult, but will rarely need the plastic measures required in the case of the post-cricoid tumours.

(iii) *Hypopharyngeal tumours.*—The hypopharyngeal tumours are still more difficult to treat and give an even poorer response to treatment than do the foregoing. Their response to distant irradiation is so poor and so temporary that it can only be regarded as palliative. Removal of an early tumour on the lateral wall of the piriform fossa may be practised by the operation of lateral pharyngotomy, a block dissection of the lymph glands of the neck being completed as part of the operation. It is often possible to close the lateral pharyngeal incision but, in the case of a large tumour, it may have to be left open and packed. The growths on the posterior wall of the post-cricoid region can be removed by a lateral pharyngotomy and removal of the posterior wall of the hypopharynx and upper portion of oesophagus. A large cervical skin flap must be turned in and fixed at the site of the excision and the wound closed much later. These methods are applicable only in comparatively early growths but, as they hold out the only hope of successful removal, are better than irradiation or not attempting any form of treatment.

(iv) *Epilaryngeal or epiglottic tumours.*—Small tumours of the edge of the epiglottis have been removed successfully through the mouth by means of the epiglottic punch forceps, devised by Lake and Secombe Hett. It is only the occasional case that is circumscribed enough to be safely removed in this way Larger tumours can be removed through a lateral pharyngotomy and as there is not any loss of tissue of the wall of the pharynx the results are good. Tumours of the epiglottis which have extended too far to be removed by lateral pharyngotomy, and especially those which have spread into the valleculae or base of the tongue, can be removed by subhyoid pharyngotomy or by median translingual pharyngotomy. The results of irradiation of tumours in this position are disappointing.

Differentiation of cells

In deciding between surgery and irradiation the degree of differentiation of the cells as first shown by Broders may be of help. The most differentiated cells are radio-resistant and as they extend slowly are more suitable for surgery. The least differentiated cells are more radio-sensitive and spread rapidly and are, therefore, more suitable for irradiation. A hard-and-fast rule cannot be made, but these are contributory factors which should be taken into account. It can hardly be emphasized too strongly that irradiation of any type should not be undertaken without the intimate co-operation of an experienced radiotherapist and of a physicist who is well versed in the clinical application of *gamma*-rays and x-rays Without this guidance great harm can be very easily done to a patient and any benefit would be purely by chance.

(*d*) *Laryngofissure or thyrotomy*

Laryngofissure (Fig 149) consists of the sagittal division of the thyroid cartilage throughout its length, of cutting through the lining mucous membrane and of drawing apart the two halves of the larynx. The interior may then be

directly inspected and any necessary treatment carried out under local or *regional anaesthesia*; there is, however, always a certain amount of strain and shock and in the majority of cases a general anaesthetic will give the best results. As compared with general anaesthesia, local anaesthesia is more liable to be followed by reactionary haemorrhage; under regional anaesthesia also it is sometimes very difficult to stop a persistent capillary oozing.

Anaesthetic

(i) *Tracheotomy.*—A tracheotomy should be done as the first step of the operation, its object being to ensure an adequate airway throughout the operation and also to enable blood to be kept out of the trachea and bronchi. If there is no laryngeal obstruction general anaesthesia may be induced and continued throughout the operation, but when there is even a minor degree of obstruction it is better to open the trachea under local anaesthesia and to induce general anaesthesia through the tracheal cannula.

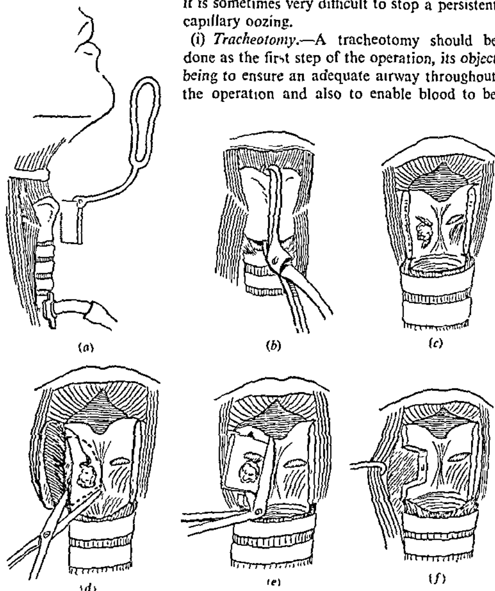


FIG 149—Stages in the operation of laryngofissure (*a*) Making a groove in the thyroid ... (*b*) dividing the cartilage with laryngeal shears, (*c*) interior of larynx showing ...

The skin should be infiltrated with a 2 per cent solution of Novocain from

the level of the hyoid bone to the suprasternal notch and over an area about 2 inches wide.

(ii) *Incision*—The incision can be carried down to the thyroid cartilage, the cricothyroid membrane and the cricoid cartilage. The anterior wall of the trachea is cleared and, as the tracheal cannula must be far enough from the larynx so as not to incommode the operator, it is usually wise to divide the thyroid isthmus, ligating the two halves and the communicating branches of the superior thyroid arteries.

The trachea is anaesthetized by injecting ½ cubic centimetre of a 10 per cent solution of cocaine to which has been added a few drops of adrenaline, and then it is opened through the third and fourth rings and the cannula inserted. The upper part of the trachea is packed with gauze through the tracheotomy opening.

The thyroid cartilage is cleared over a narrow field of about ½ inch, but the cricothyroid membrane should be cleared over an area 1 inch wide. A little trouble may be caused by bleeding from the cricothyroid branches of the superior thyroid arteries which will require clamping and ligating. A few drops of the cocaine and adrenaline solution should be injected through the cricothyroid membrane which is then incised transversely, midway between the borders of the thyroid and cricoid cartilages. Through this opening a strip of ribbon gauze is packed down the trachea on to the gauze already inserted into it from below so that the lumen is completely shut off from the operation field in the larynx, thus preventing blood from entering the lower air passages. The interior of the larynx can then be packed with narrow ribbon gauze soaked in the cocaine and adrenaline solution for anaesthetic and haemostatic purposes.

Opening of the larynx

The thyroid cartilage is incised along the full length of its midline, but in men over 40 years of age it is usually ossified and a specially shaped saw is used to cut a gutter in the cartilage. This should not pass right through the cartilage but should be deep enough to engage the blade of the shears which are used to divide the remainder of the cartilage and mucous membrane. Great care must be taken to keep this groove in the sagittal plane and this may be done more conveniently by the surgeon moving to the head end of the patient and sawing from above.

The gauze pack is removed from the interior of the larynx and the long curved blade of the shears introduced gently through the opening in the cricothyroid membrane and passed up between the vocal folds until it can be felt immediately above the notch in the upper margin of the thyroid cartilage. The shears are then closed very firmly and on lifting them out the larynx is opened along the anterior commissure. The two halves of the thyroid cartilage are drawn or held apart by hooked retractors, a mastoid retractor or Killian's long-bladed nasal speculum. The interior of the larynx can now be inspected and palpated and the appropriate treatment carried out.

Excision of epithelioma

Excision of an epithelioma of a vocal fold is the most common aim of this operation. After opening the larynx, inspecting the growth and planning how far the soft tissues need be removed to obtain a safe margin of healthy tissue, a periosteal elevator is used to lift the muco-perichondrium off the inner face of the thyroid wing over its entire surface; the muco-perichondrium should be stripped off the cricothyroid membrane and off even the cricoid cartilage

if necessary to get well below the growth. The perichondrium, carrying the attachments of the thyroid group of muscles, is separated from the outer face of the thyroid cartilage, which now stands inside a perichondrial space, as the septal cartilage stands isolated at the comparable stage of a nasal submucous resection. The thyroid ala is removed either by using a Ballenger swivel-knife, as used in the septum operation, or by clipping it out by means of a pair of scissors. The advantage of removing the cartilage is that it gives much more room for the next stage of operation, and it also prevents necrosis and sequestration of the cartilage after the operation. The soft tissues are next cut through with a straight pair of scissors, one cut being made through the upper part of the vestibular fold, the other being as low in the subglottic region as possible. The mass of soft tissue carrying the growth is now only attached posteriorly to the arytenoid process and adjacent cricoid cartilage. It is held gently by its anterior margin and separated by using a special pair of scissors bent on the flat at a right angle. These scissors are pressed as far back as possible and will sever the mass of soft tissue in one cut, which often includes the vocal process of the arytenoid cartilage. There is always a little oozing from the posterior cut which may require ligation of a small vessel, packing with gauze soaked in cocaine and adrenaline, or the application of a little snake-venom styptic powder. It is very important to obtain complete haemostasis, since, if this is not done, blood will trickle down into the lower airways and cause inflammatory conditions in the lungs. As soon as satisfactory haemostasis has been effected the gauze plugging is removed from the trachea, retractors are removed, and the wound is closed by suturing the perichondrium and soft tissues together; then, after dusting a little penicillin and sulphathiazole powder into the wounds, the skin incision is closed with interrupted sutures or clips.

Removal of cartilage

Freeing of tumour

Removal of tumour

Haemostasis

(iii) *After-treatment.*—The patient is returned to bed and placed at once in a sitting position. The tracheotomy tube should be left in position for a period of 6–24 hours, but not longer. It ensures a clear airway, easy respiration and encourages haemostasis It can be removed when the patient has completely recovered from the anaesthetic and is able to cough up any secretion at will and able to make his needs and desires known.

Removal of tracheal cannula

For the first 6 hours or so the mouth may be wiped out and moistened, but after that small sips of water may be taken, and after 24 hours ordinary soft diet may be given.

Mild sedatives may be given during the first night, but need only take the form of aspirin, with phenacetin and a little codeine. It is advisable to let the patient sit out of bed for a few minutes on the afternoon of the day after operation and to get up for increasing periods afterwards. The wound, which needs to be covered only with a strip of gauze and some collodion, heals by first intention and the sutures or clips may be removed on the fifth or sixth day. The tracheotomy wound takes up to 10 days to heal, but needs no special attention except to keep it clear of collected secretion.

Healing

The interior of the larynx heals over by granulation tissue and takes from 6 to 8 weeks for this to happen. A bar of fibrous tissue forms and acts as an immobile vocal fold, towards which the opposite fold over-adducts and a voice is produced.

In the case of the child with multiple papillomas the same procedures are

carried out with the exception that all the tumours are carefully clipped off the mucous membrane and nothing else is removed.

Webs and stenosis

In the case of webs and fibrous stenosis, following injuries, healed ulceration and too high a tracheotomy, the larynx is opened in the way described and the excess fibrous tissue carefully dissected off. There will be a considerable area without epithelium and adhesions may probably result. A mould of the interior of the larynx is made in stent, and is so constructed that a projecting portion passes out through the cricothyroid membrane and has a flange which fits the skin accurately. This flange can be kept in position by adhesive strapping. The obturator part of this mould is covered with a Thiersch skin graft, so that the raw surface of the graft is in contact with the raw surfaces in the larynx. The obturator is left unmoved for 10 days or more and may then be gently withdrawn, cleaned and replaced. It will be necessary to continue the use of the obturator for 6–8 weeks before the interior of the larynx is sufficiently epithelialized and the risk of further stenosis has passed.

Stent

(e) *Laryngectomy*

Laryngectomy (Fig 150) means total removal of the larynx. It may be done by opening the hypopharynx and dissecting the larynx out downwards—the Gluck-Sorensen method; alternatively it may be removed by cutting across the trachea immediately below the cricoid cartilage and dissecting the larynx upwards, finally separating it by cutting round the pharyngeal tube. This is known as the operation of Perier.

T-shaped incision

(i) *Incisions.*—The incision may be T-shaped with the transverse part lying just below the hyoid bone and parallel to it, and the vertical part lying in the midline and reaching to a point just above the suprasternal notch. If there is any reason to suspect metastases in the cervical lymph glands and a block dissection is intended, a second transverse incision is made at the lower end of the vertical limb and then two lateral flaps can be dissected up For simple laryngectomy these lateral incisions are unnecessary as the T-shaped incision gives sufficient access for the removal of the larynx. The difficulties with regard to drainage in a T-shaped incision have been removed by the use of a penicillin and sulphonamide powder.

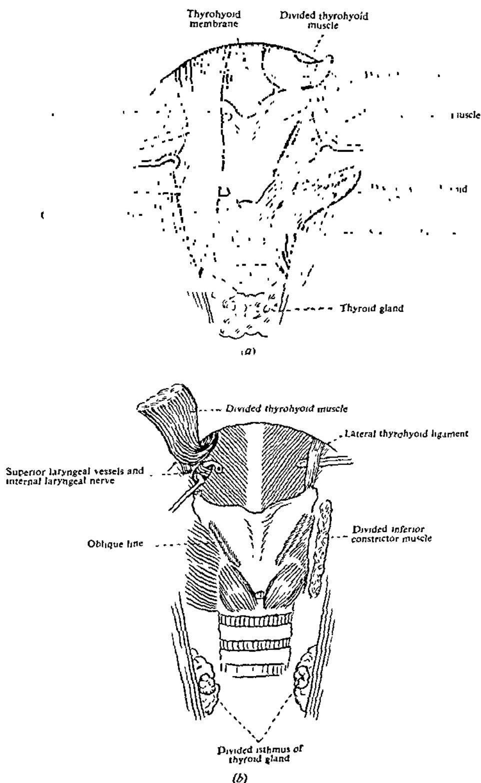
U-shaped incision

An alternative incision is a U-shaped one, with a base marked by the line of the hyoid bone and the horizontal part lying just above the suprasternal notch This incision gives very wide access and is sufficient for removal of lymph glands along the carotid sheath. Drainage, if desired, is easily obtained by introducing tubes through the lateral limbs of the incision. The flap is dissected up and can be anchored up over the chin throughout the operation. This incision is probably the best for cases in which a wide field of tissue is to be excised, but for a simple laryngectomy the T-shaped incision gives all the access necessary.

Anaesthetic

The operation is best carried out under general anaesthesia, since there is a considerable amount of manipulation in the neck and also traction on the larynx, which though not actually painful may be very uncomfortable and is productive of shock.

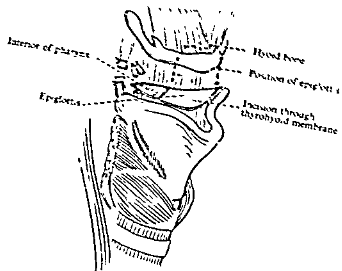
In the presence of laryngeal obstruction the skin incision should be made after infiltration with 2 per cent Novocain solution, and the upper part of the trachea and thyroid isthmus is cleared. The isthmus is divided as for



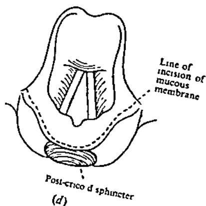
Fi[illegible] ... operation of laryngectomy. (a) Division of muscles; (b) division of ...

completed, stomach tube *in situ*, U-shaped flap to be ...

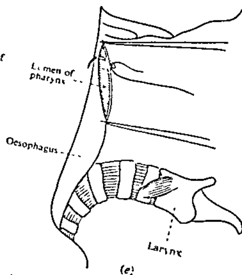
LARYNGECTOMY



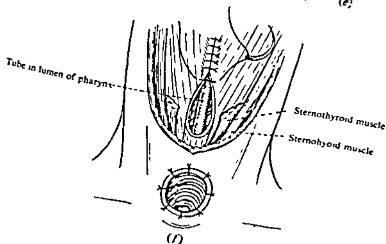
(c)



(d)



(e)



(f)

laryngofissure and a tracheotomy tube introduced. General anaesthesia may be induced and continued through the cannula. In the occasional case without obstruction, an endotracheal catheter may be introduced through the nose or mouth and the tracheotomy dispensed with.

Mobilization of larynx

The next step is the mobilization of the larynx and this is carried out by working from below upwards. The two halves of the thyroid isthmus and its lateral lobes are gently dissected away from the lateral wall of the trachea and larynx, branches of the superior thyroid artery being clamped and ligated. The sternothyroid, thyrohyoid and inferior constrictor muscles are identified and divided near their thyroid attachments. The superior laryngeal artery, vein and nerve are identified between the hyoid bone and thyroid cartilage, are ligated by passing an aneurysm needle round them, and are divided between ligatures. The superior cornu of the thyroid cartilage is freed by dividing the stylopharyngeus muscle and the lateral thyrohyoid ligament.

The thyrohyoid membrane is divided along its whole length on both sides, and the mucous membrane of the pharynx is incised immediately above the upper margin of the thyroid cartilage. The fibrous and areolar tissues between the epiglottis and hyoid bone are divided and the larynx can be grasped by a vulsellum forceps and drawn forwards. The incision through the pharyngeal mucosa is continued around the outer aspect of the aryepiglottic fold and the posterior aspect of the arytenoid process. The larynx is separated from the anterior wall of the oesophagus by gauze dissection of the areolar tissue which connects the two. The larynx can then be drawn out of the wound and is attached only by the trachea. It is finally separated by cutting through the trachea one or two rings below the cricoid cartilage. During the latter stage of the operation fouling of the wound by secretions from the pharynx should be prevented as far as possible by gauze packing.

Separation of larynx

The opening of the pharynx is closed by careful suturing. Catgut is used and the sutures should pass through the submucous tissue and muscular coat only, and should not penetrate the mucous membrane itself as the suture would favour the formation of a fistula. A rubber stomach-tube is passed through the nose, through the pharynx just before it is finally closed, and down the oesophagus to the stomach. The mucous membrane of the pharynx is turned in as the sutures are tied, and a second row of sutures, chiefly including the muscular coat, is inserted, burying the first row. The most difficult area to make secure is the uppermost part just below the hyoid bone, and this can conveniently be sutured in the shape of a T. The deep fascia and the divided muscles are sutured together and as the skin incision is closed the trachea is firmly sutured to the skin round its whole circumference and a Lombard-Moure laryngectomy tube put in position. The remaining portion of the trachea may be sutured to the skin at the lower end of the T-shaped incision or in the horizontal portion of the U-shaped incision. A useful variation is to bring the trachea to the surface through a stab incision between the lowermost part of the main incision and the upper margin of the sternum. This leaves a band of untouched skin between the operation area and the tracheostoma, which reduces the chance of infection from the wound reaching the air passages. The trachea is sutured to the skin edge by a number of silk or silkworm-gut sutures, some of which should pass round the tracheal cartilage. The patient is nursed sitting up, is fed for 10 days through the stomach tube,

the trachea is kept free of secretion by means of an electric aspirator and the sutures are removed about the sixth day.

(f) Interstitial irradiation

The method of implanting rad um needles lateral to a tumour in the larynx was developed by Harmer and Finzi (Harmer, 1932; Morton, Gray and Neary, 1944) (Fig. 151). A curved incision is made over the lateral aspect of the thyroid cartilage. The sternohyoid muscle is retracted and the sternothyroid and thyrohyoid muscles are separated from their attachment to the cartilage.

A quadrilateral flap of perichondrium is dissected up so that the denuded area covers the site of the growth to be treated. The cartilage in this rectangle

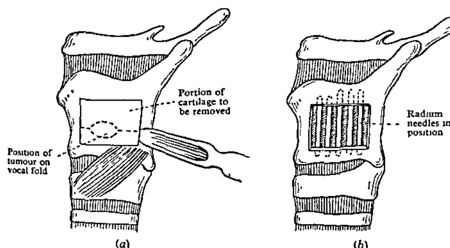


FIG. 151.—Implantation of radium needles for interstitial irradiation (Harmer-Fi
(a) Fenestration of thyroid ala; (b) needles in position.

is removed by scraping it away with a gouge or spoon curette. Care should be taken not to perforate the perichondrium on the inner aspect of the cartilage. At the anterior end of the window are introduced two needles each containing 1·3 milligrams of radium and, posterior to these, three needles each containing 2 milligrams of radium. The needles are slipped through the window and fixed in a vertical palisade, with their two ends between the cartilage and the perichondrium The silk ligatures may be left attached in order to facilitate removal, which is done after 5 days. There is a certain amount of reaction which takes the form of a swelling of the vestibular fold and which does not finally disappear for 3 months. During this period the growth on the vocal fold flattens out and disappears, usually within 4 or 6 weeks. The voice may return to normal, but in many cases there is a little residual hoarseness.

Reaction

(g) Lateral pharyngotomy

The lateral approach to the epilaryngeal and hypopharyngeal region is known as the transthyroid pharyngotomy, and includes removal not only of part of the wing of the thyroid cartilage but also of the greater cornu of the hyoid bone (Fig. 152).

A median tracheotomy is performed as a first step. If it is anticipated that

skin flaps will be required, the incision for the tracheotomy should be transverse instead of the usual vertical one.

Incisions An incision is made along the anterior border of the sternomastoid from a point opposite to the angle of the jaw to a little below the level of the cricoid cartilage. A second incision is made from the upper end of the first one

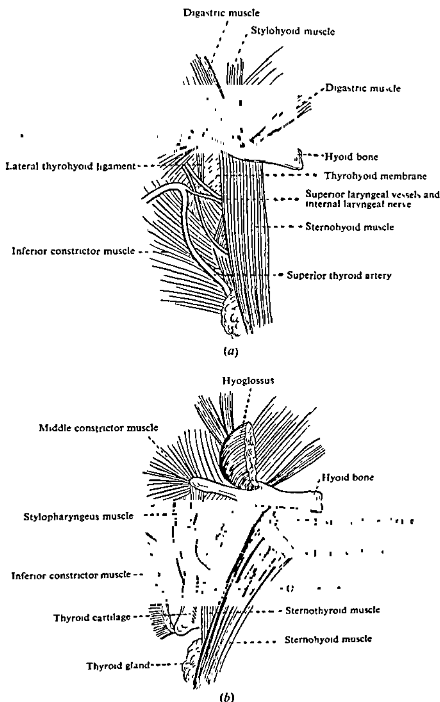
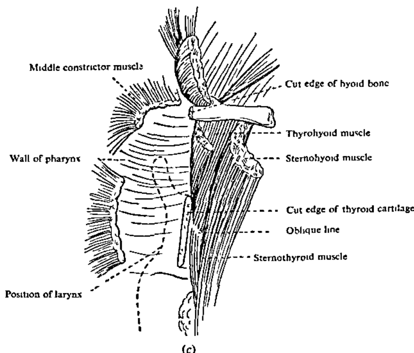


FIG. 152—Stages in the operation of lateral pharyngotomy. (*a*) *Lateral relations of pharynx*, (*b*) exposure of wall of pharynx and division of sternohyoid, hyoglossus and inferior constrictor muscles; (*c*) excision of portions of hyoid bone and thyroid cartilage; (*d*) pharynx opened, exposing the larynx.



(c)

transversely at the level of the hyoid bone to the midline. In many cases dissection of the cervical lymph system will be necessary and will be carried out at this stage. The omohyoid and sternohyoid and the muscles attached to the thyroid cartilage are divided The perichondrium is dissected up on the outer aspect of the thyroid wing and as far forwards on the inner aspect as possible. The greater part of the cartilage is removed by cutting through it with strong scissors and the greater cornu of the hyoid bone is separated from the body and removed. The lateral wall of the pharynx is cleared, and palpation will give much information concerning the extent and position of the growth. If the growth is deemed to be operable, the incision through the pharyngeal wall may be planned as part of the act of excision and will be so made as to clear the margin of the growth by ½ inch, if possible. After careful examination of the interior of the pharynx, the incision is extended to include the growth if on the lateral wall. If on the epiglottis, the growth can be removed and the opening into the pharynx closed. If on the lateral wall, and if the excision has not been too extensive, the incision may be closed, otherwise it is better left open, packed with gauze and closed by a plastic operation at a later date.


(d)

Post-cricoid growths may be removed together with that part of the pharyngeal wall which is involved, and *it will be* necessary in order to cover the resulting defect to turn in large flaps of skin which are hinged anteriorly or posteriorly according to whether the growth is on the anterior or posterior wall of the post-cricoid tube. A further plastic operation will be necessary later to close the resulting fistula.

The tracheotomy tube should be retained for a few days and a *feeding tube* reaching to the stomach left in position until the pharyngeal wall is completely healed.

(*h*) *Translingual pharyngotomy*

For tumours which involve not only the epiglottis but also the vallecula and the posterior third of the tongue, the operative measures described do not [illegible]

[illegible]

and the splitting of the tongue along the midline until the tumour is reached.

Preliminary tracheotomy

A preliminary tracheotomy is performed. An incision is made in the midline dividing the lower lip and reaching the mandible to its lower margin. It is then continued downwards on to the thyroid cartilage. The deep fascia is cut through as far as the hyoid bone, the mylohyoid muscle is divided vertically through the raphe if present—otherwise through the muscular fibres. The two geniohyoid muscles are separated and the mandible is then divided in the midline with a fine saw. If it has not already been done it may be necessary to remove one or both lower central incisors. After packing off the opening of the larynx with a small sponge or a pad with a silk thread attached, the tongue is split very accurately in the midline until the tumour mass becomes palpable.

Splitting of tongue

The incision is then carried laterally to either side of the mass. As the tongue is split backwards the halves of the mandible can be drawn apart. The triangular portion of the posterior part of the tongue can be drawn first to one side and then to the other and the incision carried round into the outer part of the vallecula, across the aryepiglottic folds and finally across the base of the epiglottis. The hyoid bone is cut through on both sides of the body, and this central portion is removed with the tumour mass.

Closure of wounds

The two halves of the tongue and the floor of the mouth are sutured together very accurately and firmly, particular attention being paid to the part immediately adjacent to the root of the epiglottis. This is facilitated by the absence of the central portion of the hyoid bone, allowing the two halves to come together more readily. The lip and skin over the chin are sutured and a drain is left in the submental region. The mandible is held in position by the soft tissues and, as a rule, does not require fixation. Food is given by a stomach tube for a week or so until swallowing can be carried out comfortably and without disturbing the mandible. The tracheotomy tube should be retained until all bleeding has ceased.

Irradiation

It may be that better results can be obtained with irradiation by radium or high-voltage x-rays than by these operations of pharyngotomy. The irradiation of these tumours is a matter of great difficulty and should only be attempted under the guidance of a radiotherapist and a physicist.

5. PARALYSIS OF THE RECURRENT LARYNGEAL NERVE

Recurrent nerve paralysis

Paralysis of the inferior or recurrent laryngeal nerve is characterized by immobility of the corresponding vocal fold and arytenoid process. The vocal fold remains motionless in the midline or in a few degrees of abduction, which is the position of rest. Either one or both nerves may be involved and their implication may be simultaneous or at an interval. Unilateral paralysis is commoner than bilateral paralysis in the proportion of 3 or more to 1. The paralysis may be caused by malignant tumours of the upper third of the oesophagus or of the thyroid gland, or the nerves may be injured during thyroidectomy or implicated in scar tissue as a result of this or other operations in the neck.

Fibrosis

Fibrosis at the dome of the pleura may implicate the nerve on either side. On the left side the nerve may be involved in the intrathoracic part of its course by tumours of the oesophagus, bronchus, lung, and mediastinum, and by dilatation of the aorta and of the left auricle of the heart. There are many cases of paralysis of one—either right or left—or of both vocal folds for which a cause is never found. In a limited number of them, partial or complete recovery takes place and the cause may be some form of peripheral neuritis.

In cases of bilateral paralysis the voice is not of necessity much altered, especially when the folds are in the midline position as they are in continuous apposition and can be set in vibration by the expired air. The narrow glottic space is, however, barely wide enough for respiration and some degree of dyspnoea is always present. This is greater when the paralysis comes on rapidly and when the two folds become immobile at the same time. In such conditions the dyspnoea is usually severe and acute, and requires tracheotomy as an urgent measure. If the paralysis comes on more slowly, and especially if it affects one side at a later date, the dyspnoea is neither so acute nor so complete, but a tracheotomy usually becomes necessary eventually.

Treatment

Speech therapy

Treatment consists in removal of the cause or repair of the injury whenever this is possible. Occasionally this may be possible in the case of unilateral paralysis, but in most patients nothing can be done to renew the activity of the nerve. In these circumstances the assistance of a speech therapist is most valuable to increase the power and range of the mobile vocal fold. Attempts have been made to renew the neuromuscular function by anastomosis of the proximal end of a divided phrenic nerve to the distal end of the recurrent nerve and have produced some successes. The phrenic nerve must be divided at the inlet of the thorax to enable its proximal end to be carried behind the carotid sheath to reach the recurrent nerve beyond the usual point of injury. Paralysis of the diaphragm will result, but if that portion which arises from the roots of the third and fourth cervical nerves only is used, recovery of movement will occur after some months.

Other methods of relieving the dyspnoea are planned to separate the cords permanently. Attempts have been made to move the anterior attachments of either one or both vocal folds outwards, or of one upwards and the other downwards. These have not given any measure of success.

More recent and more successful operations have been devised by King and by Kelly in America.

King's operation

King's operation consists of a lateral approach to the larynx through the neck (King, 1939; Seed, 1942). The pharyngeal mucosa is dissected off the thyroid cartilage, out of the piriform fossa and off the posterior aspect of the cricoid and arytenoid cartilages. The crico-arytenoid joint is almost completely disarticulated; the arytenoid cartilage is drawn outwards and, by means of a suture passed round it, is secured to the thyroid cartilage. A second ligature is passed round the arytenoid cartilage and through the divided anterior belly of the omohyoid muscle. These two ligatures contrive to hold the arytenoid cartilage in an abducted position and, by a large knot suitably placed on the first of the ligatures and fixed at the anterior border of the arytenoid cartilage, outward rotation can also be achieved. An alternative method is to pass the first ligature twice round the arytenoid cartilage and, by pulling on one end of the ligature, outward rotation can be made before the ligature is tied.

Kelly's operation

Kelly's operation consists in exposing the thyroid ala, and carrying out a window resection of the postero-inferior portion of the cartilage, the identification of the position of which is facilitated by a colleague passing a laryngoscope and transilluminating the thyroid cartilage from within (Kelly, 1941; McCall and Gardiner, 1943). The arytenoid cartilage throws a shadow and its position is, therefore, fixed. The cartilage is reached by cutting away the inner layer of the perichondrium and then working through the internal thyro-arytenoid muscle. It is held by a specially designed hook and may be drawn out of the window just made. It may then be removed, suturing the cut end of the internal thyro-arytenoid muscle. A modification of this method consists in leaving the arytenoid fixed in the window and its attached muscles sutured to the external periosteum.

These two operations separate the posterior ends of the vocal folds sufficiently for the purpose of ordinary respiration. The voice though rough is usually adequate.

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[References to other titles are given under Larynx—Diseases of, in the Index Volume. The subject is also dealt with under the heading of Larynx Diseases in the *British Encyclopaedia of Medical Practice* (1938), Vol. 7, p. 612]

LAW IN RELATION TO SURGERY

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1. THE SURGEON'S QUALIFICATIONS

217.] "Every person who enters a learned profession", said Tindal, C.J., in *Lanphier* v. *Phipos*[1], "undertakes to bring to the exercise of it a reasonable degree of care and skill. He does not undertake, if he is an attorney, that at all events you shall gain your case; nor does a surgeon undertake that he will perform a cure; nor does he undertake to use the highest possible degree of skill. There may be persons who have higher education and greater advantages than he has; but he undertakes to bring a fair, reasonable and competent degree of skill". This oft-repeated dictum still holds good as a statement of the law. We shall have to consider first what degree of skill is necessary to constitute the surgeon's qualifications.

Degree of skill

[1] (1838) 8 C. & P. 475 at p. 479.

There is a widespread belief that once on the Medical Register, a medical man is *ipso facto* qualified to carry out any diagnostic or therapeutic operation, however complex or highly specialized it may be. This, however, is not the case, and no one, whether on or off the Medical Register, is entitled to undertake any operation for the performance of which he has not acquired the requisite experience and skill. The misconception is largely based upon another, namely, that the principal object of the Medical Acts is to protect the medical man. In fact, the purpose of these Acts is to benefit the public and is expressed in the brief preamble to the Medical Act of 1858, which is as follows: "Whereas it is expedient that persons requiring medical aid should be enabled to distinguish qualified from unqualified practitioners: be it therefore enacted . . ." There is indeed no reason in law why unregistered practitioners should not undertake major surgery and, provided that they can show the requisite experience, skill and care, successfully defend any action for damage they may cause. There is, however, this difference; that in the case of a registered practitioner the requisite degree of skill will be presumed until the contrary is proved, whereas in the case of an unregistered practitioner there is no such presumption, and the onus of proof of skill is on the practitioner. It may well be, however, that so long as the unregistered practitioner does not represent himself as being other than what he is, the degree of skill required of him may be less than that required of a registered practitioner, so that the former might successfully defend an action for damages in circumstances in which the latter might fail to do so. We are for the moment concerned only with the question of skill. There are, of course, a few statutory restrictions on the activities of unqualified persons, for example, under the Venereal Diseases Act of 1936; but in general the law is impartial in its requirements as to the practitioner's skill, whether he is on the Register or not

Purpose of the Medical Acts

Requisite qualification

The question as to whether a surgeon has or has not the requisite qualification to perform a given operation is one not of law but of fact. The possession of a higher qualification in surgery such as a Fellowship of the Royal College of Surgeons, will not of itself necessarily qualify a surgeon to perform any operation. Thus, to take an example, a Fellow of one of the Royal Colleges of Surgeons who has been employed in an administrative capacity for a number of years and who decides to retire into a country general practice in which he can realize his youthful ambition and "do a bit of surgery" could scarcely, without proper further training and experience, be considered as competent to operate. On the other hand, a higher qualification in surgery would be weighty evidence of the requisite training in cases in which the possessor had made surgery his career.

Emergencies

We have already seen that a surgeon does not undertake to bring to his work the *highest* degree of skill. The degree of skill to be expected of a surgeon to a remote cottage hospital would not be that which would be expected from a surgeon to a teaching hospital in, say, London or Manchester. So also if, in a genuine emergency, a general practitioner using all possible care attempts an operation for which he has not the requisite experience and skill, he will not be responsible for any damage that may result. However, the general practitioner's excursions into operative and manipulative fields of which he has not competent experience should be reserved for such emergencies, and he should

not lose sight of the fact that the entry of his name on the Medical Register does not give him *carte blanche* to undertake any surgical procedure he pleases.

2. THE LEGAL DUTIES OWED BY THE SURGEON TO HIS PATIENT

(1) Contractual duties

The surgeon may, in the first place, by express agreement or contract undertake certain duties to his patient. Thus he may agree for a specified fee, or even without the fee being actually specified, to operate and subsequently personally to perform certain services such as dressings or removal of sutures. Such a contract, although not in writing, would be enforceable and an action would lie for its breach.

(2) Duty of skill and care

Quite independently of contract, however, the law casts upon the surgeon a duty of skill and care[1]; nor is it necessary to aver an express contract so long as it is alleged that the surgeon was retained and entered on the case[2]. The duty to have competent skill has already been considered. As regards the duty of care, this has been said to be threefold: first, a duty to exercise care in deciding whether to undertake the case; secondly, a duty to exercise care in deciding what treatment to give; and thirdly, a duty to exercise care in carrying out the treatment[3]. So a surgeon who, having either no experience or not sufficient experience in cranial surgery, undertakes, for example, a difficult intracranial operation, fails in his duty under the first of these heads, even though the operation is the correct form of treatment and though in the actual performance thereof he uses the utmost care of which he is capable. As regards the degree of care, the law demands that this shall be "reasonable". Having regard to the peculiar relationship of surgeon and patient reasonable care must be interpreted as being a high standard of care. It is probably a correct statement of general principle to say that a surgeon must exercise such skill as he possesses to the best of his ability. As to what has been held to be a breach of the duties of skill and care we shall have to consider in some detail in the section on *Negligence*.

Duty of care

(3) Duty not to disclose confidences

It is uncertain whether there is a legal duty upon a medical man not to disclose confidences. Scrutton, L.J., in *Tournier* v. *The National Provincial Bank*,[4] thought that there was such a duty. In that case it was held that a bank was under an obligation of secrecy concerning its client's affairs, and the learned Lord Justice stated that this obligation probably applied also to solicitors and doctors. The question is of particular importance to the surgeon in connexion with the provision of certificates and reports on patients. The safe course is to regard all case-notes and other medical records of patients, skiagrams, pathological reports and the like as falling within the duty under consideration, and

[1] *Pippin* v. *Sheppard* (1822) 11 Price 400.
[2] *Gladwell* v. *Steggal* (1839) 5 Bing. N C. 733
[3] 22 Halsbury's Laws 318; *R.* v. *Bateman*, 41 T L R. 557.
[4] [1924] 1 K.B. 461.

Patient's consent

in no case should such records and reports be disclosed without the patient's consent to any person not actually concerned in the treatment of the patient. However, in hospital practice it is probable that the term, persons concerned in the treatment, may be construed rather widely and may include certain lay officers, such as almoners. Even when records are required as evidence in court, the patient's consent should first be obtained, but when such consent is unreasonably withheld the Judge will order their production if they are a necessary part of the evidence. It should be noted that any *duty* not to disclose confidences does not confer a *privilege* to withhold information when it is required by the Court in evidence. It is in the discretion of the Judge as to whether or not information to which a patient objects shall be given in evidence. A doctor who refuses to give evidence when required by the Court to do so may be committed to prison.

A medical man treating a case of venereal disease under the National Scheme may be compelled to disclose the fact in evidence, notwithstanding that the statutory regulations for the scheme enjoin absolute secrecy[1].

Disclosure in evidence

Apart from the legal duty to disclose in evidence, a medical man may be under a duty as a citizen to disclose information obtained in a professional capacity. Thus it is his duty to assist in the detection and arrest of anyone committing a serious crime. Accordingly, if he attends a person who, to his knowledge, has committed a felony he himself commits an offence if he fails to inform the proper authority[2].

It is difficult to define the limits of this duty. Bankes, L.J., in *Weld-Blundell* v. *Stephens*[3] said that there was not a duty to disclose every completed wrong. He instanced the case of a man charged with attempted suicide who confesses his guilt to his solicitor; in this case the learned Lord Justice thought the solicitor would be justified in respecting the confidence. On the other hand he thought that when a patient expressed his intention to commit suicide to his doctor the latter might be under a duty to disclose the confidence. What of the case in which a doctor knows that some injury is threatened to a third party? Thus a surgeon may know that a man who is about to marry has active venereal disease. Assuming that the patient refuses to abandon or postpone his plan, is the surgeon justified in informing the lady?—or her father? When the injury to the third party would involve the commission of a criminal offence there is no doubt that the surgeon would not only be justified but would be under a duty to do all he could to prevent it. But it was held in *R. v. Clarence*[4] that the communication of venereal disease to a wife is not an assault occasioning bodily harm within the meaning of the Statute if the wife consents to intercourse. There are, however, some grounds for believing that a medical man may be released from his obligation of secrecy on the ground of social or moral duty. In the law of defamation these grounds, in the absence of malice, are a defence and there is a tendency to apply the rules governing privilege in defamation to professional confidences. On the whole, therefore, it seems likely that a surgeon would be justified in making the communication in the problem under discussion to the person to whom he would be under a social or moral duty to disclose it, and this would probably include both the lady and her father.

...ial or ...al duty

[1] *Garner* v. *Garner* [1920] 36 T.L.R. 196. [2] 1 Hale 372. [3] [1919] 1 K B 520
[4] (1889) 22 Q.B D 23.

(4) Duty to obtain patient's consent to physical interference

It is the duty of a medical man to obtain the consent of a patient to any physical interference. If he fails to do so, he is guilty of an assault (or more correctly, a battery). It is reasonable that a patient should be told what is about to be done to him that he may take courage and put himself in such a situation as to enable him to undergo the operation[1]. Thus the doctor who adroitly incises a whitlow for a nervous patient before the patient is aware of his intention to do a cutting operation may be acting with great skill and psychological insight but none the less he has committed a battery. However, consent need not necessarily be expressed. It will readily be implied from the patient's behaviour. So, if the patient sees the doctor take up his scalpel and makes no protest, this will be evidence of implied consent. Further, in an emergency, consent will also be implied, as when the patient is unconscious. So also, consent will be implied when during an operation an emergency arises and an unexpected mutilation, as for example amputation of a limb, has to be carried out to save life. More difficult, however, are the cases in which a surgeon obtains consent to an operation in the course of which, in the absence of any sudden emergency, he encounters some indication to exceed the consent given. This happens particularly in cases of neoplasm in which at operation the growth is unexpectedly found to be malignant. Many surgeons make it a rule in cases of breast tumour to ask for consent to a radical amputation if this should be found necessary, however innocent the tumour may seem. This is, of course, throwing an additional and often unnecessary anxiety on the patient, but nevertheless it is desirable to take this precaution. Supposing, however, that the surgeon has failed to seek such consent and that he does find that the growth is malignant, what is he to do? Undoubtedly, if he proceeds to radical amputation he is taking some risk of legal proceedings, and amputation of the breast is such a serious mutilation in a woman that the sympathy of a jury might well be with the patient. This is the more so because at the present time there are usually alternative methods of treatment which the patient might have elected to have had if opportunity had been given. On the whole, the surgeon who finds himself in such an unfortunate position would be well advised to close the wound and to seek consent for the further operation. It may be remarked that the consent of the husband or other near relative who may be accessible during the operation will not replace the consent of the patient herself, although it could doubtless be urged as a mitigating circumstance. So, also, a husband has no right to refuse consent to an operation on his wife to which she herself has consented[2]. When at operation a surgeon is faced with the necessity to take unforeseen steps entailing less serious results to the patient—apart from life-saving measures which as has been indicated he will always be entitled, indeed obliged, to take—he will have to decide each case on its merits. The principle involved was stated thus by Hawkins, J., in *Beatty* v. *Cullingworth*[3]: "If a medical man with a desire to do his best for his patient undertook an operation he should have thought it was a humane thing to do everything in his power to remove the mischief provided that he had no absolute definite instructions not to

Implied consent

Consent for further operation

[1] *Slater* v. *Baker & Stapleton* (1767) 2 Wils. (C.P.) 359.
[2] *Harris* v. *Lee* (1718) 3 P. Wms. 484.
[3] *The Times*, Aug. 11th, Nov 17th, 18th, 1896, and Jan 14th, 1897.

operate [*Note*: operate here meaning to extend the operation in an unexpected direction]. Would any one of them undertake an operation fettered by such a condition as the plaintiff suggested, namely, that if the [extension of the] operation was in their opinion absolutely necessary still they should not perform it if they found there was a little more disease than they had thought?" This seems to mean that if a surgeon considers it absolutely necessary to do a little more than he intended, consent to do so will be implied. It is difficult to suggest what may be included under the expression "a little more". In *Beatty* v. *Cullingworth* (*supra*) the surgeon expected to find only one ovary diseased. At operation, however, both were found to be diseased and he removed both. Immediately before the operation the patient, who was a nurse, had said, "If you find both ovaries diseased you must remove neither"; to which the surgeon replied, "You must really leave that to me, Nurse. I know your wishes and you may be sure that I shall remove nothing that I can help". The patient made no further protest. The jury found in favour of the surgeon and the Court of Appeal refused an application for a new trial. It is indeed difficult to see what other verdict the jury could have found, for the surgeon had surely made it clear on what terms he was prepared to undertake the operation, while the patient still had an opportunity to refuse the operation on those terms.

Extension of operation

It is important that the surgeon should not assume any false confidence as a result of the decision just referred to. When embarking upon any operation in which bilateral oöphorectomy or orchidectomy is even remotely likely to be necessary, this should be carefully explained to the patient and his or her consent obtained to do as much as may be necessary. Further, although it is much open to question[1] whether it is necessary in law, it is very desirable that in the case of a married woman the husband's permission should be obtained. Husbands aggrieved in this way may be a source of great trouble to the medical man responsible, and, curiously enough, they will usually be supported in their complaints by the patient who has herself consented to the operation.

Oophorectomy or orchidectomy

Consent by infants

When an infant (that is, a person under 21 years of age) resides with a parent or guardian, it is advisable to obtain the consent of the parent or guardian to an operation. Undoubtedly an infant can give valid consent to an operation if he has attained the age of discretion, but *prima facie* the consent of the father should be obtained. It is impossible to say with certainty in what circumstances the father's consent will be excused, but as the matter does not appear ever to have been litigated it cannot be of great practical importance. Probably all that is required is that the surgeon should act reasonably and in the best interests of the patient.

Consent of the father

(5) Consent and criminal liability

So far we have discussed the question of consent to operation from the point of view only of the civil liability of the surgeon, that is, the liability of the surgeon to compensate the patient by paying him damages. Apart from this, however, the surgeon who operates on a patient without consent, or without valid consent, may also find himself liable to a criminal prosecution. We may dismiss immediately all those cases in which there is a proper indication for surgery. In such cases, although a technical battery may have been committed

[1] See the views of Atkin, L.J., in *Tr. of Medico-legal Soc.*, 1925, XIX, p. 93.

the law will leave the patient to his civil remedy in damages. When, on [illegible]

consent had not even been sought. What are the cases which come within this rule? In his *Digest of the Criminal Law* (6th ed.) Art. 227, Sir James Fitz-James Stephen enunciates the proposition that "every one has a right to consent to the infliction upon himself of bodily harm not amounting to a maim"[1]. Of this proposition Swift, J., said in the Court of Appeal in *R.* v. *Donovan*[2]: "This may have been true in early times when the law of the country showed remarkable leniency towards crimes of personal violence but it is a statement which now needs considerable qualification." In Donovan's case the appellant had been convicted of a common assault upon a girl whom, for perverted sexual gratification, he had beaten with a cane. The defence was consent. The Court of Appeal took the view that "if the jury were satisfied that the blows struck by the prisoner were likely or intended to do bodily harm to the prosecutrix they ought to convict him" and that the question of consent need not even be considered. Further, the meaning of "bodily harm" was there defined as "its ordinary meaning and includes any hurt or injury calculated to interfere with the health or comfort of the prosecutor. Such hurt or injury need not be permanent, but must, no doubt, be more than merely transient or trifling". It would seem likely, therefore, that anyone who performs an operation which has as its object the ovarian sterilization of the patient on grounds merely of convenience and in the absence of some pathological indication, commits a criminal offence punishable by imprisonment. Indeed it is highly probable that the mere surgery of access, involving as it does the major operation of opening the abdominal cavity, would in itself be within the above definition of "bodily harm".

Bodily harm

The only other class of case in which the surgeon may be asked to operate in the absence of any indication on the ground of disease or deformity is that in which some beautifying or cosmetic operation is sought, as in face-lifting. According to Lord Riddell (*Medico-Legal Problems*, 1929) the legality of some of these operations has been questioned, although the same author concluded that the law seems to be that such operations are not unlawful since they are not maiming operations. Neither do they come clearly within the definition of bodily harm. Lord Riddell, however, poses some interesting questions; for example, is a wife entitled to disfigure herself as she thinks fit? If the beautifying operation turns out badly, can the husband sue the surgeon? Since the object of the operation is to beautify and not to disfigure, the first question scarcely arises, and as to the second question, so long as the operation itself is lawful and is performed with the patient's consent, it is submitted that an action could be founded only on negligence.

Cosmetic operation

3. NEGLIGENCE

Negligence is the breach of a duty owed by one person to another. Without some existing duty there can be no negligence. Thus, if a surgeon sees the

[1] A maim is such an injury as would interfere with a person's fighting efficiency and the term certainly includes castration.
[2] 25 Cr. App. Rep. 1.

victim of a street accident bleeding to death in the road and callously refrains from lending assistance, he commits no tort, for he owes the victim no duty. But let him once offer his assistance and actually attend to the patient and he will immediately owe him the duties of skill and care which we have already discussed. Malpractice or malpraxis is another term which is sometimes used to signify negligence. If negligence be found the surgeon can claim no fee and is liable in damages for the loss and suffering caused to the patient. Such damages are recovered in a *civil* action for negligence.

Malpractice

Criminal negligence.—When an injury has been caused by negligence of a particularly gross kind the surgeon may be criminally liable and if death has resulted he may be prosecuted for manslaughter[1]. It is not possible to define with any precision the degree of negligence that will be regarded as necessarily criminal. Various epithets have been used by judges, such as "gross", "culpable" or "wicked", but perhaps it is best defined as "such disregard for the life and safety of others as to demand punishment". The delicate line has to be drawn by the jury.

The authorities are clear that in a prosecution for criminal negligence the question whether the accused was or was not a duly qualified practitioner is not in itself material in determining his guilt[2]. "If a person bona fide and honestly exercising his best skill to cure a patient performs an operation which causes the patient's death, he is not guilty of manslaughter. . . . It would be most dangerous for it to get abroad that if an operation performed either by a licensed or an unlicensed surgeon should fail, that surgeon would be liable to be prosecuted for manslaughter." (*Per* Hullock, B., in *R.* v. *Van Butchell*[3].)

Civil actions for negligence

Prosecutions of surgeons for negligence are excessively rare. Civil actions, on the other hand, are not infrequent. Most commonly the question is raised by way of counter-claim in an action by the surgeon to recover unpaid fees. If the patient intends to defend the action he adds little to his costs by raising the counter-claim whereas by so doing there is always the chance that he will succeed, thereby evading payment of the fees and getting damages and costs into the bargain.

Apart from these often trumpery claims there is the really important group of cases in which the patient has, or believes he has, a genuine claim for damage suffered as a result of something the surgeon has done or has failed to do. These cases may be classified under the following heads:

(1) Negligence in diagnosis.
(2) Negligence in selecting treatment.
(3) Negligence at operation.
(4) Negligence in after-treatment.

(1) Negligence in diagnosis

We have already considered the duties of skill and care, the breach of which constitutes negligence. We have now to consider in relation to diagnosis how far the surgeon may be rendered liable for mistake. It would appear that there is a difference in the liability of the surgeon according to whether he has undertaken the care or treatment of the case or whether he is simply examining

[1] *R.* v. *Bateman* [1925] 41 T.L.R., 557; 19 Cr. App. Rep. 8.
[2] 1 Hale 429.
[3] [1928] 3 C. & P. 635.

the patient for the purpose of making a report to another person. Thus in *Pimm* v. *Roper*[1] a surgeon was employed by a railway company to examine a passenger who had been injured in a collision. He formed the opinion that the patient "was more frightened than hurt", and acting on this opinion the patient accepted £5 by way of compensation from the railway company. The next day a hernia was discovered. The patient had not called attention to his abdomen and the surgeon did not examine it. The passenger unsuccessfully sued the railway company for this further alleged injury. He then sued the surgeon. The surgeon was held not liable, there being no proof of injury on account of the surgeon's neglect to examine the abdomen. This case was cited with approval by Scrutton, L.J., in *Everett* v. *Griffith*[2] who, speaking of the implied undertaking of the doctor to his patient, expressed the principle as follows: "But it appears to me the case is quite different when the doctor is not operating on or treating the patient, but expressing an opinion about his condition at the instance of another person and to guide that person in independent action." In such cases the undertaking (to the patient) to bring the appropriate degree of care and skill is not implied, but "submission to a surgical operation or medical treatment is a good consideration on which to found this undertaking".

Making a report

It should not be assumed that a surgeon will never owe a duty of care and skill to the patient when he is retained by a third person for the purpose of making a diagnosis only. Indeed, in most cases quite the opposite is true. It is submitted that the test is, "was the surgeon retained to act for the benefit of the patient?" If the answer is "yes", as when the surgeon is consulted for the purpose of an independent opinion, then the duties will be implied, and it makes no difference that the surgeon was retained by a husband or wife, an employer or a charitably minded total outsider. If the answer is "no", then the su geon will not be liable to the patient for a mistake in diagnosis, for such an action could be founded only on a breach of the implied duties referred to above.

Assuming that the case is one in which the duties exist, the question as to whether the mistake arose from a breach of one or both of them is one of fact for the jury, to be decided on the evidence. A surgeon having the requisite skill, using all care and doing his best for his patient cannot be made liable for a mistake in diagnosis, however serious are the consequences.

Mistake in diagnosis

Failure to take skiagrams

An allegation that the surgeon has been negligent in failing to take skiagrams for the purpose of diagnosis or treatment is only too frequent in practice, and the defence societies urge that skiagrams should always be taken in all cases of injury to bony parts. Whether failure to do so will amount to negligence will depend upon the facts in any case, but it cannot be too strongly emphasized that such stress has been laid on the desirability of this rule from both medical and medico-legal quarters in recent years that it would be difficult indeed to excuse a failure to take skiagrams in any case in which there has been a danger of bone injury.

The writer has not been able to find any reported English case in which the question of failure to take skiagrams has been a main issue, although the

[1] (1862) 2 F. & F. 783. [2] [1920] 3 K.B. 163 at p. 193.

question has repeatedly been considered in unreported cases. In the Canadian case of *Moore & Moore* v. *Large*[1] it was held not necessarily negligent to omit to take skiagrams, even if the diagnosis turned out to be erroneous, and in the Ceylon case of *Subapathi* v. *Huntley*,[2] a motor accident case, the decision was similar.

(2) Negligence in selecting method of treatment

The matters that require discussion here are (a) errors of judgement, (b) unorthodox treatment, (c) novel treatment and (d) operations of exceptional danger.

(a) *Errors of judgement*

As to errors of judgement, it will be clear from the general principles determining liability for negligence that only when a breach of the duty of skill and care can be proved will such liability exist. In other words, if a surgeon, having the requisite skill and using all due care, makes an honest error of judgement in selecting the treatment, he will not be liable however disastrous the result may be.

(b) *Unorthodox treatment*

Reasonable method of treatment

As to unorthodox treatment, here liability for damage that may result will depend upon whether the method of treatment can be shown to be reasonable, that is, reasonable according to present-day standards of surgery. Thus a totally obsolete method would not pass this test. It is difficult to say at what stage of obsolescence a surgical procedure becomes legally unjustifiable. Suppose that a given radical operation for cancer is statistically proved to produce inferior results and a much higher mortality rate than non-operative radium treatment does, is the occasional diehard surgeon failing in his duty to the patient if, being honestly unconvinced of the superiority of the new treatment, he continues to operate? Is the barbarous method of treating simple facial naevi in infants by carbon dioxide snow, which is still occasionally carried out, legally justifiable in view of the almost perfect curative and cosmetic results which can be obtained by irradiation? Perhaps some day a jury will have to decide these questions, and much may depend upon how the evidence on each side is presented. On the question of alternative methods of treatment an interesting Canadian case is *Kinney* v. *Lockwood Clinic*[3] in which it was laid down that a surgeon is liable for negligence when he minimizes the risk of an operation in order to induce his patient to undergo it, and fails to explain a possible alternative treatment.

(c) *Novel treatment*

As to novel, that is, untried or insufficiently tried, treatments a learned judge once said, in *Slater* v. *Baker & Stapleton*[4]: "... For anything that appears to this Court this was the first experiment made with this new instrument and if it was it was a rash action." In that case a distinguished surgeon to St. Bartholomew's Hospital had to pay heavy damages to a patient for whom he had broken and reset a badly united fracture, applying traction extension by means of "a heavy steel thing with teeth" . ' trument referr

[1] [1932] 4 D L.R. 793. [2] [1938] 1 W. [3] [1931] 4
[4] (1767) 2 Wils

judge. A further ground of liability was his failure to obtain the patient's consent. Perhaps the surgeon's real fault here was that he was in advance of his time. Fortunately, the courts now take a much more liberal view, requiring only that the treatment must be reasonable and well grounded, and that the nature of the treatment and its attendant risks should be explained to the patient.

(d) Operations of exceptional danger

As to operations of exceptional danger it is clear once again that the surgeon is under a duty to explain the danger to the patient. Is there any point at which an operation becomes too dangerous to be justifiable even with consent? In the early stages of development of very extensive radical operations, such as oesophagectomy, the mortality rate may be very high indeed. Is such an operation justifiable? Clearly all the circumstances of the case would have to be considered. Such an operation would probably be justifiable in the case of a patient incurable by any other means and faced with the prospect of certain death from his disease, provided that there could be said to be a real chance of cure or marked alleviation. So also, such an operation might be justifiable in an effort to relieve continuous and unbearable pain incurable in any other way. No definite rules, however, can be laid down for these cases, and indeed it is clear that any rigid rule would necessarily tend to hamper the development of surgical technique.

Chance of cure

(3) Negligence at operation

The law regards an operation as team-work, each member of the team having his allotted task, and the surgeon is entitled to rely on the members of his team for the performance of their individual tasks. This rule is, however, not unqualified. In the first place, it is the duty of the surgeon to select competent persons to form the team and if he fails in this duty he (or the hospital[1]) will be liable. Secondly, it is the duty of the surgeon to see that there is a reliable system of working in the theatre. Thus, for example, the system of swab-checking should be as foolproof as it is possible to make it. The anaesthetic arrangements must also be such as to ensure, as far as is humanly possible, against the risk of explosion, for example, in cases in which electrical sparking is used Thirdly, in assessing liability all the circumstances of the case will be taken into account. So, in the course of a critical and difficult operation for perforated duodenal ulcer, a surgeon cannot be expected to count the swabs himself after removing all of which he is aware (*Mahon* v. *Osborne*[2],) provided that a satisfactory system of counting swabs by some other member or members of the team is proved. In that case three months after the operation the patient became acutely ill. A second operation disclosed a swab between the under surface of the liver and the stomach, as a result of which an abscess had formed. The patient died the next day. In an action under Lord Campbell's Act and the Law Reform (Miscellaneous Provisions) Act, 1934[3], a verdict was found for the plaintiff but the Court of Appeal (Scott, MacKinnon and Goddard, LL.J.) ordered a new trial on the ground of misdirection of the jury.

[1] As to the incidence of responsibility on hospital and surgeon respectively, *see* p 388.
[2] [1939] 2 K.B. 14.
[3] *See* page 394.

Mahon v. *Osborne* is an important case because it clearly lays down certain rules of law which are stated below; but before stating them it is necessary to explain briefly the doctrine known as *res ipsa loquitur*. Ordinarily, when negligence is alleged the burden of proving the negligence is upon him who alleges it; that is to say, it is for him to adduce evidence which will satisfy the court that the defendant has been negligent. In certain cases, however, the damage complained of is so obviously due to negligence that "the thing speaks for itself". The classical case is *Byrne* v. *Boadle*[1] in which a barrel of flour rolled out of a warehouse window on to a passer-by. The doctrine applied simply because "properly handled barrels do not behave in this way". The importance of the doctrine is that when it applies, the onus of proof is shifted to the defendant who must then prove he was *not* negligent. In other words, a presumption of negligence is raised which the defendant must rebut or else lose the case. This is a much more serious matter for the defendant than merely waiting for the plaintiff to produce what evidence of negligence he can and then attempting to demolish the plaintiff's case. In *Mahon* v. *Osborne* (*supra*) the Court of Appeal held (Scott, L.J., dissenting) that in the case of a swab left in the body the doctrine of *res ipsa loquitur* applies. The Court also held, as we have seen, that there is no general rule of law requiring a surgeon at the end of an operation, after removing all swabs of which he is aware, to make sure that none are left in the patient's body.

Res ipsa loquitur

Onus of proof

It must not be assumed from the above decision that as a general rule a surgeon is absolved from counting swabs. The circumstances of that particular operation were such as to necessitate all possible speed in closing the abdomen, as well as complete freedom for the surgeon to concentrate on the main task. When, however, the operation is a simple one the surgeon will have greater difficulty in absolving himself. So in *Dryden* v. *Surrey C.C. & Stewart*[2] a surgeon was held liable for leaving a plug of gauze in the patient, as a result of which she sustained a renal infection.

The doctrine of *res ipsa loquitur* has been held to apply in a Canadian case (*Hugheston* v. *Jost*[3]) in which an intravenous injection was allow to escape into the tissues, and in another Canadian case (*Cox* v. *Saskatoon*[4]) in which an injury was sustained during a transfusion. However, in the English case of *Morris* v. *Winsbury-White*[5] the doctrine of *res ipsa loquitur* was held not to apply when part of a rubber tube was left in the bladder at, or at some unknown time after, prostatectomy. The changing of tubes was carried out by various members of the hospital staff who, it was held, in the performance of their duties were not under the control of the surgeon and he therefore could not be fixed with liability under the *res ipsa* doctrine for an act of negligence which might have been the act of any one of a number of persons.

Inevitable accident

This has been defined as "an accident not avoidable by any such precautions as a reasonable man doing such an act then and there could be expected to take". Accident falling within this definition is a good defence to an action for negligence. Thus in *Gerber* v. *Pines*[6] a doctor was held not liable for negligence when a needle accidentally broke in a patient's arm, part being left

[1] (1863) 2 H & C. 722. [2] [1936] 2 All E.R. 535. [3] [1943] 1 D.L.R. 402.
[4] [1942] 1 D.L.R. 74. [5] [1937] 4 All E.R. 494. [6] (1934) 79 Sol. Jo. 13.

behind in the tissues. It was also held in that case, however, that the doctor was negligent in not informing the patient of the accident, and the patient was awarded five guineas damages, but no costs, on this ground. This case shows that a patient is entitled to be informed of any accident of the sort, and such information should be conveyed as soon as possible.

Inform the patient

(4) Negligence in after-care

Claims under this head arise most frequently in such lesions as Volkmann's contracture, plaster sores and bedsores. The general principles of liability, already sufficiently discussed, will apply.

Questions frequently arise as to how far the surgeon may entrust the after-care of patients to house-surgeons and nurses. In *Morris* v. *Winsbury-White* (*ubi-supra*) Tucker, J. (as he then was), dealing with a case of prostatectomy said: "In the normal case . . . the obligation [of the surgeon] was to pay such visits as were necessary after the operation to see how the patient was progressing under the care of the nurses in the hospital and the resident medical officers there"; and in that case the duties of changing and shortening bladder and perineal drainage tubes were held to be properly entrusted to nurses and resident medical officers. In general, the rule is that a surgeon may entrust to other members of the staff such duties as may be reasonable and as they are qualified to undertake. "Reasonable" here means reasonable as judged by the standards of surgeons in general, not necessarily what one particular surgeon thinks reasonable. When such duties have been properly entrusted the surgeon will not be liable for any acts of negligence on the part of those carrying them out: "I think it well established as a matter of law that the resident medical officers in a hospital of this kind and the nursing staff are not the agents of the specialist surgeon who comes and performs an operation of this kind." (*Per* Tucker, J., in *Morris* v. *Winsbury-White* (*ubi-supra*).) In the same case it was also held that when a surgeon contractually undertakes to give a patient his personal attention this does not necessarily imply that he will himself attend to such matters as the changing of drainage tubes.

Obligation of the surgeon

It is probable that the same consideration would apply to private nursing homes as to hospitals. This, however, is by no means certain and it is possible that a nurse assisting at an operation or carrying out specialized work for a surgeon may be regarded as temporarily passing under the control of the surgeon. It will be noted that the dicta in Morris's case are confined to hospitals "of this kind" (St. Paul's Hospital, a public voluntary hospital) and are, therefore, not wide enough necessarily to cover nursing homes. When, however, the duties are entrusted to persons who are actually employed by the surgeon himself, for example, a consulting-room nurse, then the surgeon will be liable for the acts of negligence of his employee.

Contributory negligence of patient

If the surgeon can establish that the patient could probably have avoided the damage in respect of which he is suing had he himself exercised reasonable care, he may now have this taken into account in the assessment of damages. Prior to the Law Reform (Contributory Negligence) Act, 1945, contributory negligence of the defendant was an absolute bar to the recovery of damages.

Allied to the question of contributory negligence is the duty of the patient

Mitigating the damage

to do all he can to mitigate the damage. If he fails to do so, as in rashly and unreasonably failing to follow advice in after-treatment, this also may be taken into account in assessing damages.

4. HOSPITAL SURGEONS

When a patient enters a hospital for treatment, the hospital undertakes certain duties to him. So also does every member of the hospital staff who ministers to his care whether that person be surgeon or nurse, wardmaid or porter, and a breach of duty by any of these parties will found an action for negligence against the defaulting party. The liability of these various parties is, however, not mutually exclusive. To some extent liability overlaps. Our first task, therefore, is to ascertain as exactly as possible the respective liability of the surgeon and of the hospital authorities for the acts of each other and of the hospital staff.

We may take as a starting-point the proposition that a master or principal is liable for the wrongful acts of his servant or agent:

(1) When they are the consequence of his own specific orders.

(2) When they are committed "in the course of his employment" or "within the scope of his authority".

The first branch of this rule needs no elaboration. The second branch depends upon the principle that a person who puts another in his place to do a class of acts, necessarily trusts him for the manner in which the acts are done; and the liability will not be limited to the acts which are expressly within the servant's or agent's authority but will also extend to acts which are within his implied authority, for example, some unusual action for the benefit of his employer or principal in an emergency.

Servants and agents

We next have to inquire who are servants and agents? Is a surgeon a servant or an agent of his hospital? Or a nurse? Is a nurse in that relation to a surgeon when acting under his orders? Or a house-surgeon? Or an anaesthetist?

It has long been settled that a visiting physician, having freedom to use his independent judgement in the treatment of his cases, is not the servant or agent of the hospital, which will accordingly not be liable for his negligence (*Evans* v. *Liverpool Corporation*[1]). Until 1942 however, as a result of the well-known decision in *Hillyer* v. *St. Bartholomew's Hospital*[2], the position of a nurse was peculiar inasmuch as she was regarded, for some purposes, as the servant of the hospital and for other purposes as an independent professional person in the same legal position as the visiting physician. Thus if a nurse scalded a patient while serving him with his tea—a "ministerial" or non-professional duty—the hospital would have been liable since for that purpose she was the servant of the hospital. If, on the other hand, she scalded him with an over-hot fomentation, that is, in the course of a "professional" duty, the hospital would not have been liable. However, in 1942 the Court of Appeal in the important case of *Gold* v. *Essex County Council*[3] fully reconsidered the law with regard to the responsibility of hospital authorities, doctors, nurses and others employed by them and put forward new principles which they applied in that case, and which probably now govern the law in these matters. The legal principles were stated by MacKinnon, L.J., as follows.

[1] [1906] 1 K.B 6. [2] [1909] 2 K.B. 820.
[3] [1942] 2 K B 293; [1942] 2 All E.R. 237.

(1) One who employs a servant is liable to another person if the servant does an act, within the scope of his employment, so negligently as to injure that other.

(2) That principle applies even though the work which the servant is employed to do is of a skilful or technical character, as to the method of performing which the employer is himself ignorant; for example, a ship-owner and a certified captain who navigates his ship.

(3) The liability of the master for the negligent act of the servant will exist although at the time the servant is by direction of the master, or by operation of law, under the control of a third party; for example, when the captain of a ship is under the command of a Naval Commodore in charge of a convoy.

(4) The master will not be liable for the act of his servant if he is only doing without personal negligence that which he is directed to do by such third party. That is to say that if a nurse did precisely and without negligence on her part what she was ordered to do by a surgeon, the hospital would not be liable, but that if she did negligently that which she was ordered to do by the surgeon then the employer would be liable.

Contract of service

Contract for services

Lord Justice Goddard in the same case distinguished between a contract *of service* and a contract *for services*. A contract of service is one in which the employee works under instructions either from the employer or from some other person appointed by the employer. We have seen that for the negligent carrying out of such instructions the employer is liable. On the other hand a contract for services is one in which the person employed to carry out the services is not subject to control in carrying them out, but is entitled to act as he thinks fit in any part cular circumstances. It should be noted that a contract of either class is not necessarily a formal or even a written contract. An oral appointment implies a contract of the one or the other class. The obvious example of a contract for services is that of a visiting physician or surgeon attached to a voluntary hospital. Such an officer is subject to no control in connexion with his professional work, and accordingly for his negligence the hospital authority will not be liable. On the other hand it was held in Gold's case that the contract of a radiographer (that is, a technical assistant and not to be confused with a radiologist) was one of service and the defendants were held liable for his negligence. So also the contract of a nurse is one of service, and the rather artificial distinction between the types of service she renders is no longer of importance as regards nurses[1].

Liability of house-surgeons

As to the liability of house-surgeons, the case of *Collins* v. *Hertfordshire C.C.*[2] is of considerable importance. In that case a surgeon requested his (unqualified) house-surgeon to arrange for the preparation of 100 cubic centimetres of a 1 per cent solution of procaine for use at operation. The house-surgeon mistakenly ordered from the pharmacist 100 cubic centimetres of a 1 per cent solution of cocaine. No written prescription was given, and the pharmacist took no steps to verify this unusual order. At operation the surgeon injected the solution and the patient died as a result. In an action brought by the widow *against the Hertfordshire County Council (who maintained the hospital)* and the surgeon, it was alleged that the Council permitted

[1] But as regards medical officers, *see Lindsey C.C.* v. *Marshall (infra)*

[2] [1947] 1 All E R. 633.

a dangerous system of working, and that the patient's death resulted from the negligence of the Council in its conduct of the hospital, and that the Council was vicariously responsible for the negligence of the surgeon, the house-surgeon and the pharmacist.

Mr. Justice Hilbery found: (i) that the surgeon was negligent and was personably liable; (ii) that the Council was negligent, and was also liable for the negligence of the house-surgeon and of the pharmacist but not of the surgeon. Thus, it seems clear from this decision that a house-surgeon is in the position of a servant from the point of view of vicarious liability of the hospital, and it is submitted that it makes no difference that in this case the house-surgeon was unqualified.

In any event a hospital will be *liable for the negligence of its medical officers* in carrying out *administrative* duties. So in *Cull* v. *Chance*[1] a hospital was held liable for the performance of a hysterectomy contrary to the wishes of the patient as expressed in a letter from her general practitioner, which reached the house-surgeon but was not communicated to the surgeon. So, also, in *Lindsey County Council* v. *Marshall*[2], a House of Lords case, the defendants, who maintained a maternity home, were held liable for failure of the staff of the hospital to inform an incoming patient of the danger of puerperal sepsis, and for failing to take proper precautions for disinfection. Hillyer's case (*supra*) was distinguished on the ground that the duties were there professional and not administrative.

Negligence of subordinates

As regards the liability of surgeons for the negligence of their subordinates acting under their directions this was considered in *Perionowsky* v. *Freeman*[3] in which an action was brought against two surgeons at St. George's Hospital for alleged negligence in the treatment of a patient there, by placing him in an excessively heated bath and keeping him in it for an improper length of time whereby he was severely scalded. The surgeons were personally present at the time. The defence was that the injury was due to the negligence of the nurses. Cockburn, C.J., said that hospitals could not be expected to engage a staff of medical men to attend to all the minor incidents and details of treatment, and that medical men who gave their services gratuitously were not to be made liable for negligence for which they were not persona ly liable. This is a strong case, as the surgeons were present during the proceeding at which the patient was injured. It will be noted that the gratuitous character of the services of the surgeons was referred to by Cockburn, C.J., but it is probable that there is now no difference in the liability of the surgeon in negligence, whether his services are gratuitous or paid. The recent decision in *Morris* v. *Winsbury-White* (*supra*) supports the principle of Perionowsky's case. It should be noted that the liability of the hospital in the latter case was not considered, the action being against the surgeons only.

(1) Responsibilities in relation to anaesthetists

The anaesthetist, like any other member of the staff, will be responsible for his own negligence. It is necessary to inquire, however, how far that responsibility may be shared by others. On the principles laid down in Gold's case (*supra*) it is clear that the hospital authorities will not bo liable. How far can

[1] *The Times*, June 18th, 1932. [2] [1937] A.C. 97. [3] (1866) 4 F. & F. 977.

the surgeon for whom the anaesthetic is given be rendered liable? When the duty of selecting the anaesthetist falls upon the surgeon, he might be rendered liable for selecting a person of whose competence he did not take sufficient care to assure himself. So, also, if he allows a student or house-surgeon with little experience to administer an anaesthetic in a case requiring experienced judgement. He may also render himself liable if he disregards the anaesthetist's warnings and encourages him to give the anaesthetic in dangerous dosage, nor will his acting on the surgeon's encouragement absolve the anaesthetist from his liability. When, however, the surgeon has used all due care in the selection of the anaesthetist and has not been a knowing party to the anaesthetist's negligence, it would seem clear that he cannot be held liable, on the principle, previously referred to, that he is entitled to rely on the members of his team for their individual duties, but he is not absolved from all liability. He is not entitled, for example, to ignore danger signals due to anaesthetic overdosage or idiosyncrasy simply because he regards them as within the anaesthetist's province. These liabilities extend to external dangers in connexion with the anaesthetic, as well as to dangers associated with the administration. Thus, to allow an exposed electric fire or flame in the theatre dangerously close to an explosive anaesthetic would be negligence both on the part of the surgeon and the anaesthetist if the patient sustained damage as a result. These examples are all simple applications of the general principles of liability in negligence.

Anaesthetic overdosage

Deaths under anaesthetics.—It is the duty of the surgeon who performed the operation to notify the coroner in the case of a death under an anaesthetic.

(2) Post-mortem examinations

There is no property in a dead body in English law (*Williams* v. *Williams*[1]). There is however, a right to possession. Section 7 of the Anatomy Act, 1832, provides that any executor or other party having lawful possession of the body of any deceased person, and not being an undertaker or other party entrusted with the body for the purpose only of interment, may permit the body of such deceased person to undergo anatomical examination, unless, to the knowledge of the executor or other party, such person shall have expressed his desire, either in writing at any time during his life, or verbally in the presence of two or more witnesses during the illness whereof he died, that his body after death might not undergo such examination, or unless the surviving husband or wife, or any known relation of the deceased person, shall have required the body to be interred without such examination.

Right to possession

It will be observed that the enactment imposes no obligation on the executor or other lawful possessor of the body to consult or even to notify the near relatives but it should not be too readily assumed that there is no legal obligation to consult or to give notice to the relatives. It does seem clear, however, that if the relatives are not known, or cannot be found by reasonable inquiry, the post-mortem examination need not be dispensed with for lack of permission. As to who is the lawful possessor (in whom the right to carry out the post-mortem examination is vested subject to the provisions of the Act), it is clear, in the first place, that the executors are in this position. If there are no executors, the nearest relative is probably entitled to possession. If there are

[1] (1882) 20 Ch.D. 659.

no near relatives, the hospital is entitled to possession of the body, and in fact is the lawful possessor of the body until it is claimed or in due course interred.

(3) Property in case records and other documents

The question of the ownership of medical case records does not seem to be directly covered by authority but it has long been considered that the property is in the medical man making the records, or, in the case of a hospital, in the hospital. Strong support is lent to this view by the decision of the Court of Appeal in *Leicestershire C.C.* v. *Michael Faraday Ltd*[1]. In that case, which concerned documents prepared by a firm of surveyors (the defendants) for the purpose of making a report, MacKinnon, L.J., said: "It is a case of a professional man whom a client resorts for advice. I think it would be entirely wrong to extend to such a relation what may be the legal result of the quite different relation of principal and agent. . . . These pieces of paper as it seems to me cannot be shown to be in any sense the property of the plaintiff. . . . They are documents which he [the defendant] has prepared for his own assistance in carrying out his expert work." It would therefore seem that documents and skiagrams prepared for the purpose of giving an opinion or carrying out treatment are the property of the professional man or institution undertaking to give an opinion or to carry out the treatment. When, however, the patient goes to a radiologist for the express purpose of having skiagrams made, the position is not so clear and it may well be that in certain circumstances the skiagrams could be claimed by the patient. Mr. D. H. Kitchin[2] however, takes the view that the ownership of the films should be with the radiologist (or hospital) in the absence of an express agreement to the contrary. For a fuller discussion of this subject than is possible here the reader is referred to Mr. Kitchin's excellent work (*supra*).

Ownership of skiagrams

5. PRIVATE NURSING HOMES

It is uncertain how far the legal position of the surgeon who treats his patient in a private nursing home differs from that which obtains when he treats him in a hospital. That there are differences is very probable. In the first place, the patient's contract with the nursing home is usually to supply suitable accommodation and nursing and other services, but not medical or surgical treatment, for which he has a separate contract with the surgeon. On the other hand, a patient who enters a hospital usually, if not invariably, does so on an undertaking by the hospital, expressed or implied, to provide both treatment and other services. Accordingly there is, in the nursing home, no medical organization with resident medical officers and the like on which the surgeon can rely in his absence. The effect of this is that the whole responsibility for the control and supervision of the treatment and after-care of the patient while in the home rests on the surgeon

Responsibility rests on the surgeon

Secondly, since nursing homes vary greatly in the facilities they offer and also, unfortunately, in efficiency, there will be a duty of care upon the surgeon in his choice of a home suited to the particular needs of his patient.

Thirdly, the appointment of nurses to responsible positions in hospitals is

[1] [1941] 2 K.B 205.
[2] *Legal Problems in Medical Practice.* Arnold & Co. 1936

made by careful selection by experienced persons subject to the supervision of a committee or board, whereas the appointment of nursing-home staff is rarely subject to any such regulations and supervision. It will, therefore, usually be the duty of the surgeon to satisfy himself of the competence of any nurse to whom he wishes to entrust any responsible task.

Finally, there is the question, previously referred to, as to whether for the purposes of an operation the nurses may not become the servants or agents of the surgeon, thus throwing on him responsibility for their negligence. We have seen that it will usually be the duty of the surgeon to satisfy himself personally of the competence of any nurse to whom he wishes to entrust any responsible task. To whom belongs the duty to provide a safe system of working in the theatre? This is not entirely a nursing or an administrative responsibility, nor is the management of the average nursing home, maintained often by one or two retired nurses who undertake the supervision of every department, so intelligent or so competent as to entitle the surgeon to rely upon it for this purpose. Further, all the members of the team which provides the human side of the safe system of working are in hospital practice *provided by the hospital*, while in nursing-home practice some are *usually* provided by the surgeon. It is scarcely open to doubt that the surgeon must assure himself of the safety of the system of working, and it seems likely that he must also, as a rule, assume responsibility for the whole team in nursing-home practice. Whether for this purpose the nurses would be regarded as "servants lent", thus absolving the management from responsibility, or whether the surgeon would share the responsibility with the nursing home, would depend on the circumstances of any case.

Regulation of nursing homes

Nursing homes are defined as premises used for the nursing and care of persons suffering from any sickness or injury. They are regulated by the Public Health Act, 1936, Section 187 *et seq.*, and the Public Health (London) Act, 1936, Section 240. They include maternity homes but not government or municipal hospitals or hospitals incorporated by Royal Charter. Exemption from the provisions of the Acts may also be granted to hospitals or institutions not carried on for profit. The Acts provide for the registration of nursing homes. To carry on an unregistered nursing home is an offence punishable by fine, and in the case of a second offence, by imprisonment. Applications for registration are made to the county or county borough council. In case of refusal or cancellation of a certificate, appeal lies to a court of summary jurisdiction. The county and county borough councils are the local supervising authorities for the purposes of inspection and the like.

Registration

6. RIGHTS OF ACTION

In English law, for the breach of a contract, only the actual parties to the contract can sue. Even if a third party is damaged by the breach he cannot sue on the contract. Accordingly, if the surgeon's contract is with some person other than the patient, for example, an employer, the patient himself will not be able to sue on the contract, though if the surgeon has actually entered on the case he may, of course, be liable in tort (for example, for negligence).

(1) In third parties

Apart from the right of action in the patient himself, there are in certain cases special rights of action in third parties, and, in the case of death, rights of action devolve upon the personal representatives (that is, the executors or administrators).

(a) Husband and wife

When his wife has been injured, a husband is entitled to sue for the loss of her consortium, but only if he was in fact living with her at the time of the injury. There seems to be no reason why a wife should not also be able to sue in respect of the loss of her husband's consortium.

(b) Parent and child

A parent may sue in respect of an injury to his (or her) child by reason of which the child's services have been lost to the parent. There is no action for the loss of the child's company, or for outraged feelings, but the law will very readily presume services by the child.

(c) Master and servant

It is possible for a master to bring an action in respect of loss of a servant's services caused by the tort of a third party (*Att. General* v. *Valle-Jones*[1]).

(2) Effect of death upon rights of action

Exceptions

Since the Law Reform (Miscellaneous Provisions) Act, 1934, causes of action subsisting against or vested in the deceased at the time of his death pass to the personal representatives. There are certain exceptions, only one of which may possibly concern the subject-matter of the present article. It is provided that rights of action for defamation do not devolve, and it is possible that this may also include an action for disclosure of a professional confidence. No special time limit is prescribed for the commencement of an action for the benefit of the estate. An action against the estate, that is, if the surgeon has died, must have been pending at the date of death, or the cause of action must have arisen not more than six months earlier than the death and proceedings must be commenced not later than six months after the personal representatives have taken out representation.

When the alleged damage has been the cause of the death a separate and independent action, which may be pursued concurrently, may be based on the Fatal Accidents Act, 1846 (Lord Campbell's Act), which gives a right to compensation to the families of persons killed by torts.

It is of interest that under the former Act the personal representatives acquire precisely the rights of the deceased, and damages have been awarded for loss of expectation of life, for pain and suffering, and even for the loss of the use of a limb for a few days immediately preceding death (*Rose* v. *Ford*[2]).

(3) Limitation of actions

Special limitation periods

The law limits the period after the commission of a wrong during which an action may be brought. In general this period is six years (Limitation Act, 1939), but there are certain special limitation periods prescribed by various statutes. An action against a public authority must be brought within one

[1] [1935] 2 K.B. 209. [2] [1937] A.C. 826.

year of the alleged injury (*ibid.*). Assistant medical officers of a hospital maintained by a county council under its statutory powers are entitled to the benefit of this limitation period (*Nelson* v. *Cookson*[1]). The limitation period under the Fatal Accidents Act, 1846, is one year from the death. The limitation period under the Anatomy Act, 1832, is six months. The commencement of an action is the date of issue of the writ. The writ need not be served at once, but may be held in reserve during negotiations. Thus the non-arrival of a writ within the limitation period does not necessarily imply that none has been issued.

[References to other titles are given under Law in Relation to Surgery, in the Index Volume.]

[1] [1940] 1 K.B. 100.

LENS—DISEASES AND INJURIES

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1. CATARACT

(1) Definition

218.] The term cataract is technically applicable to any opacity of the crystalline lens. In speaking to patients it is wise to confine the use of the term to such opacities as require surgical intervention. Too often patients with non-progressive lens opacities are told that they have cataract and they worry for years in consequence.

(2) Aetiology

Congenita cataract

In most cases of cataract the cause is unknown. In a minority of cases the cause is determinable. A large group of cataracts are congenital in origin. The exact time of origin of these opacities is frequently calculable from knowledge of the architecture and development of the lens. The precise cause is seldom known. In a proportion of cases, cataract has followed an attack of German measles in the mother during the first two months of pregnancy. Other congenital cataracts have a hereditary basis.

_ndocrine dysfunction

Later in life, cataract may follow endocrine disorder. In diabetes mellitus, subcapsular lens opacities may typically occur; less characteristic cataracts of so-called senile type are also common. Cataract not infrequently follows tetany, whether from removal or disease of the parathyroids, or from rickets. The cataracts noted in dystrophia myotonica, in cretinism, in Mongolian [illegible] poikiloderma atrophicans

glass-blowers and chain-makers. It may follow exposure to lightning, to x-ray irradiation or to radium after a latent period which sometimes lasts for several years.

Cataract may follow the ingestion of poisons such as dinitrophenol in slimming tablets, and ergot, and may be produced experimentally in animals after feeding with thallium, naphthalene and galactose.

Cataract may follow perforating and non-perforating injuries of the eye. It may be secondary to iridocyclitis, to glaucoma, or to retinal lesions such as retinitis pigmentosa or to detachment of the retina (cataracta complicata).

Cataract may be caused by absorption of toxins from some septic focus such as an apical dental abscess, a tonsillar infection or a chronic appendicitis.

Cataracts in the elderly are often of dubious aetiology. Clinically they constitute the most important group and are customarily labelled senile. They are the largest single cause of blindness and are particularly important because they are generally remediable.

(3) Surgical anatomy

In cataract the lens capsule is commonly thickened and may be calcareous. The anterior capsule is composed of epithelial cells which in certain circumstances may proliferate. The posterior capsule is a hyaline membrane. The lens is supported laterally by fine strands extending to the ciliary body—the zonular fibres. The posterior convexity of the lens lies in the patellar fossa on the anterior surface of the vitreous. Sometimes the anterior surface of the vitreous is adherent to the posterior surface of the lens capsule by the so-called ligamentum hyaloideocapsulare.

A cataract is said to be incipient when it remains capable of transmitting sufficient light for useful vision. In certain cases fluid is absorbed, causing an intumescent cataract. The term "mature" is applied to a cataract when the opacity extends through the lens cortex to the subcapsular region; this term no longer has any reference to the operability of the cataract. Later the lens may shrink and dry with deposits of dense greyish-white material in the cortex, the cataract being hypermature. A late stage of disintegration in which the capsule contains a milky-white fluid, sometimes with the unliquefied nucleus in the centre, is known as a Morgagnian cataract.

(4) Pathology

Opacity of the lens takes place following disturbance of the respiration of the lens or of its nutrition. The capsule of the lens behaves as a semi-permeable membrane. Oxygen, glucose and amino acids enter the lens from the surrounding aqueous humour, while carbon dioxide, lactic acid and other excretory products pass through the capsule from lens to aqueous. There is no vascular supply to the lens except in the embryo. Oxygen is utilized in the lens by means of autoxidative systems which employ glutathione, *beta*-crystallin, and ascorbic acid. In cataract the oxygen consumption of the lens is lessened and the glutathione and ascorbic acid contents are diminished. The β-crystallin is broken down by proteases into its constituent amino acids. These latter at first attract water by osmosis from the aqueous; later they tend to diffuse out of the lens capsule, when the water content of the lens decreases. In the normal lens there is approximately 65 per cent of water by weight. This may increase to 70 per cent in sclerosis, or to 75 per cent in early

Sodium content

cataract, though the percentage decreases in hypermaturity. The sodium content of a tissue is thought to run proportionately to the extracellular water of that tissue, while potassium fluctuates in proportion to the intracellular water. This conception lends some interest to the chemical analyses of normal and of cataractous lenses. The incineration of a normal lens produces 0·5–1·0 per cent of its weight in ash. In cataract the ash may reach 1 6 per cent. Sodium in the normal lens represents 11 per cent of the ash; but 16–21 per cent is present in cataract, in which extracellular water is probably increased. Conversely, potassium decreases in the cataractous lens. There is an increase in the calcium content of the ash from cataractous lenses.

(5) Histology

The study of microscopical changes in the lens presents difficulty in avoiding artefacts during fixation. The following seem to be the chief changes. The capsular epithelium shows degenerative and proliferative changes. The epithelium may grow posteriorly to line the posterior lens capsule. Cells from this source and from the equatorial region may swell up and enlarge to form bladder cells (Wedl's vesicle cells), which frequently wander into the lens cortex. In incipient cataract spaces appear between the lens lamellae, often near the equator in the first instance. These spaces are at first filled with clear fluid; then the fluid becomes albuminous. Later the albuminous deposits run together to form Morgagnian, or myelin, globules. Later still the lens fibres themselves become opaque and eventually break down into Morgagnian globules. Brownish-red pigment may be deposited.

(6) Clinical picture

The main symptom is a gradual diminution of vision. If the opacity is mainly nuclear the patient may see better in dull light when the pupil is dilated. Lens striae may cause doubling of images, as when the points of a crescent moon may seem bifid. Some opacities may be seen entoptically as spots. Even when visual acuity is grossly diminished it is usually possible for the patient to indicate the direction of lights by pointing to them (light projection). Inaccurate projection of light is generally a sign of retinal or choroidal damage and may contra-indicate operation. An exception occurs in the case of calcareous lenses, in which proper projection may be prevented by chalky plaques. In such cases it is often possible to obtain useful vision by operation even when projection is poor.

Doubling of images

Light projection

Upon inspection by transmitted light, lens opacities stand out black against the red reflex from the fundus. Oblique illumination is often deceptive, for the lenses of many elderly people appear yellowish-grey when examined by this method. In later stages a cataract may look staringly white. Light thrown on the eye from one side will sometimes throw a shadow of the pupillary margin upon the lens nucleus. This shadow disappears when sclerosis increases, the lens being then termed mature. The eye should be carefully examined for such features as increased tension, hypotony, or posterior synechiae, which might suggest secondary or complicated cataract.

(7) Special aids to diagnosis

Use of the slit-lamp

Important information in the diagnosis of cataract may be obtained by the use of the slit-lamp. By employing this, it is possible to inspect lens opacities

under high magnification, discriminating between water clefts which may conceivably reabsorb, and irreversible lens opacities in which there has been protein coagulation. In the case of actual opacities it is possible to discriminate between progressive types and non-progressive forms of opacity, such as the various forms of congenital cataract, or the club-shaped coronary opacities often seen in the middle or deeper layers of the lens cortex. In the case of complicated cataract associated with retinal or choroidal lesions it is often possible to suspect the latter from their association with polychromatic lustre of the posterior lens cortex. Accuracy in determining prognosis is thus much enhanced by the use of this instrument.

(8) Differential diagnosis

Care should be taken to make an ophthalmoscopic examination of all cases of suspected cataract. All too frequently cases of chronic glaucoma are mistakenly diagnosed as cataract because of the gradual diminution of vision. Ophthalmoscopic examination would have disclosed cupping of the disc before the condition was allowed to progress to irreversible optic atrophy.

Chronic glaucoma

A large haemorrhage into the vitreous sometimes simulates cataract. On inspection it is impossible to get an ophthalmoscopic view of the fundus: there is a typical black fundus reflex. The lens is seen to be unaffected on slit-lamp examination.

Haemorrhage into vitreous

Difficulty sometimes arises when lens opacities are associated with retinal lesions such as senile macular degeneration or diabetic retinopathy. These conditions make for faulty light projection.

(9) Prognosis

Once a lens opacity has passed the water-cleft stage and has become a protein coagulum, the condition is irreversible. So far it has been found impossible to control the progression of lens opacities by medicinal means, although clinical experience suggests that attention to associated factors, such as diabetes or focal sepsis, may slow down their advance.

The prognosis of operative intervention is generally speaking excellent, particularly if skilled ophthalmic nursing is available.

The patient's age is in itself no contra-indication to operation, although in the case of octogenarians and nonagenarians the surgeon must remember more than ever to treat the patient as well as the local condition. In extreme old age recovery depends to a great extent upon the maintenance of a high morale in the patient. Hence the operation should be performed as a one-stage procedure rather than in several stages, even though the latter might appear to be technically the safer course.

Age of patient

(10) Indications for surgical intervention

Surgery should usually be undertaken as soon as reading becomes difficult in the case of educated patients, or, in the case of the illiterate, as soon as they find difficulty in getting about. Operation is slightly simpler if the lens is mature in the sense of being opaque throughout, but modern technique has rendered cataract extraction practicable in any stage of the condition, so that the primary consideration is nowadays the convenience of the patient. Other concurrent maladies such as arteriosclerosis, bronchitis and prostatic obstruction, are not absolute contra-indications to operation, although they may

render prognosis less favourable. Care should be exercised in choosing a period for operation when these complications are in a quiescent stage. In the case of diabetes it is important that the disease should be under careful control at the time of operation.

Unilateral cataract

There has been considerable discussion as to whether unilateral cataracts should be submitted to operation. In such cases the surgeon should carefully consider whether he thinks the expectation of life is such that the second lens is likely to become severely affected. If he thinks this likely, operation is indicated, as the general condition is likely to deteriorate, rendering the operation a more severe procedure by the time the second eye is affected. Such a patient will be delighted later on that one eye has already been dealt with. In these days of traffic congestion the enlargement of visual field obtained by operation on a unilateral cataract is advantageous. Generally speaking also the morale of a patient with unilateral cataract is higher than that of a completely blind person: this is in itself a powerful aid to a successful result, since the patient is likely to be less difficult on the operating table.

Such patients should be warned that owing to the high prismatic effect of a cataract lens, it is impracticable to use their operated and unoperated eyes concurrently. This difficulty can be overcome by contact lenses, but few old people can cope with their management.

(11) Pre-operative management of the patient

Infected teeth should be extracted. Chest, urine and prostate should be examined. Inquiry should be made concerning the patient's consumption of alcohol so as to be forewarned of any possibility of delirium tremens. A conjunctival smear should be taken to determine the nature of any conjunctival organisms present. Pneumococci, *staphylococcus aureus* and streptococci are usually contra-indications to operation. The patency of the nasolacrimal duct should be determined by syringing. The lashes should be cut short. Most surgeons prefer the pupil dilated by atropine before operation. Pre-operative sedation with phenobarbitone, grain 1, helps a nervous patient.

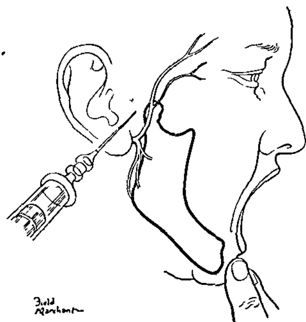


FIG 153.—O'Brien facial block.

(12) Operative technique

(a) *Anaesthesia*

O'Brien method of facial nerve block

In operations for cataract, anaesthesia is one of the most important factors in securing a successful result. Akinesia is best secured by the O'Brien method of facial nerve block (Fig. 153). The patient's mouth is opened widely and an injection of about 2 cubic centimetres

of 5 per cent procaine and adrenaline is made on to the condyle of the mandible. A small amount of procaine is injected superficially in the same region. By this manoeuvre the upper division of the facial nerve is blocked. After the operation the upper lid must be kept closed by a stitch which may be attached to the cheek by a piece of strapping. A small amount of procaine injected at the site of the stitch obviates pain on its insertion (Fig. 154). An alternative method of securing akinesia is to inject procaine deeply along the outer and lower orbital margins. Pain may be avoided by a deep retrobulbar procaine injection in the region of the ciliary ganglion on the outer side of the optic nerve (Fig. 155) An injection of procaine into the superior rectus permits a bridle suture to be inserted painlessly (Fig. 156). Superficial anaesthesia is attained by the instillation of 4 per cent cocaine drops.

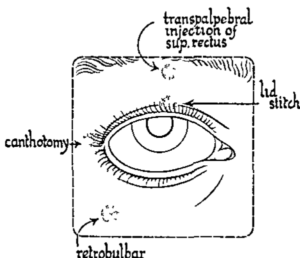


FIG 154 —Sites of supplementary procaine injections

(*b*) *Types of operation*

Exigencies of space make it necessary to describe these in outline only.

(i) *Discission.*—This consists in incision of the anterior lens capsule with a small Ziegler knife

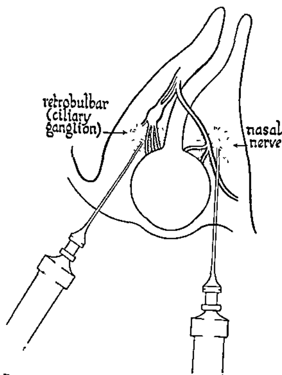


FIG. 155 —Plan showing injection of procaine near the ciliary ganglion. A block of the naso-ciliary nerve is sometimes performed also.

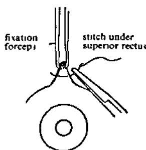


FIG. 156 —Insertion of bridle suture under superior rectus.

(Fig. 157). The contained lens matter is broken down by proteolytic enzymes and is absorbed. Sometimes the lens matter swells up and the ocular tension is raised. The lens matter then has to be evacuated by a linear extraction. The pupil is kept well dilated by atropine until all lens matter is absorbed.

Discission is best suited to cataracts in patients under 35 years of age.

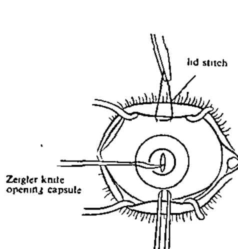


FIG. 157.—Discission

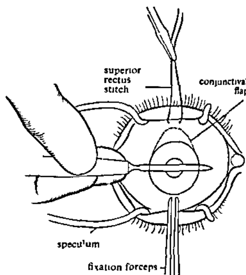


FIG. 158.—Making the "section" in cataract extraction.

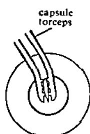


FIG. 159.—Use of capsule forceps

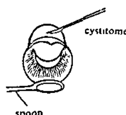


FIG. 160.—Expression of lens

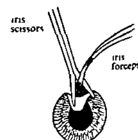


FIG. 161.—Iridectomy.

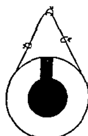


FIG. 162.—Conjunctival flap sutured in position.

(ii) *Extracapsular extraction.*—A transfixion flap is made with a Graefe knife. The flap involves nearly all the upper half of the cornea and usually is prolonged so as to raise a conjunctival flap in addition (Fig. 158). The anterior lens capsule is incised or a segment is torn out by capsule forceps (Fig. 159). The patient is made to look down and the lens is expressed from its capsule by gentle pressure just below the lower corneal margin (Fig. 160). An iridectomy is performed if the pupil does not contract readily on being replaced by an iris repositor after the lens expression (Fig. 161). The flap is sutured back into place by conjunctival sutures or, if preferred, by a corneo-scleral suture (Fig. 162). The lid is closed by a stitch attached by strapping to the cheek. The eyelids are covered by a piece of Vaseline gauze and protected by a pad and bandage.

A capsulotomy may be necessary at a later date.

(iii) *Intracapsular extraction.*—In this operation the corneal flap is cut as in extracapsular extraction, except that the section must be made a little larger. The usual procedure is to seize the anterior capsule just below the anterior pole by a pair of Arruga's forceps (Fig. 163). The zonule is then ruptured by very slow movement of the lens medially, laterally, downwards and finally upwards towards the section (Fig. 164) The lens is delivered in the capsule by gentle traction (Fig. 165). The iris is replaced by a repositor, an iridectomy

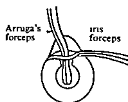


Fig 163 —Seizing the lens capsule with Arruga's forceps in intracapsular extraction.

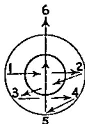


Fig. 164.—Diagram of manipulations in intracapsular extraction.

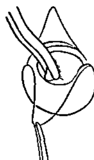


Fig 165 —Delivery of lens

being performed if the pupil fails to contract readily. The flap is fixed in place by conjunctival or corneo-scleral suture.

Intracapsular extraction has the merit of completing the operation in one procedure. It can be performed at any stage of lens opacity. The eye usually settles down more quickly than after the extracapsular method. The procedure is slightly more difficult than the extracapsular technique.

(13) Post-operative care up to convalescence

It is necessary to cover only one eye after cataract extraction, although excessive movement of the eyes may be lessened by hanging a gauze flap over the unoperated eye for the first 2 days. The main danger period is at and just after the first 24 hours, when the anterior chamber has re-formed but the wound is as yet only lightly united. Straining at this period may force an iris prolapse which may require abscission or replacement. The pupil is kept dilated with atropine until the first dressing, after 48 hours. Trauma is prevented by careful nursing supervision. Infection is rare; if it occurs, penicillin drops, 2,000 units per cubic centimetre, may sometimes save the eye. Temporary glasses are supplied in 14 days and permanent ones at about the second month.

Main danger period

Results of operation.—In properly chosen cases under 5 per cent of cataract operations are unsuccessful; and useful vision can be confidently expected if there is no abnormality of the fundus oculi.

2. CONGENITAL ABNORMALITIES OF THE LENS

In addition to congenital cataract, the lens may be notched at one side. It may also be subluxated or even dislocated into the anterior chamber of the

vitreous. Dislocations may be associated with arachnodactyly. *If vision is* affected the lens is best removed by discission as described. Secondary glaucoma is a dangerous complication (*see* Glaucoma, Vol. 4, p. 319).

Secondary glaucoma

3. EXFOLIATION OF THE LENS CAPSULE

This is a senile condition in which the superficial layers of the anterior lens capsule tend to flake off, particularly in the areas rubbed by the iris during pupillary excursions. Flakes may settle in the anterior chamber or on the zonule or may block the angle of the anterior chamber, producing a raised tension (glaucoma capsulare of Vogt). The treatment is as for chronic glaucoma.

4. INJURIES

Non-perforating injuries

The lens may sustain a non-perforating or a perforating injury. In the former case a Vossius ring may form on the anterior capsule or a cataract may develop. The lens may be extruded subconjunctivally through a subconjunctival scleral rupture, and may require removal after coagulation by surgical diathermy. It may be dislocated into the anterior chamber, whence it is best removed by a section as for cataract extraction. If dislocated into the vitreous, it is best left alone unless secondary glaucoma necessitates a vectis removal through a cataract incision. A perforating injury usually causes swelling of the lens when lens debris may have to be removed by linear extraction. Rarely a foreign body retained in the lens may become encapsulated, but the usual result is an opacity of the whole lens. Iron and steel fragments should be removed early by electromagnet to prevent the onset of siderosis oculi.

Perforating injuries

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[References to other titles are given under Lens—Diseases and Injuries, in the Index Volume. The subjects of Diseases and Injuries of Lens are also dealt with under the headings of Blindness and Cataract in the *British Encyclopaedia of Medical Practice* (1936), Vol. 2, p. 407 and (1937), Vol. 3, p 1.]

LEPROSY

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1. DEFINITION

219.] Leprosy is caused by the growth of *Mycobacterium leprae* or *Bacillus leprae* (Hansen, 1874 and 1880) in the human body and the reaction of the tissues to its presence.

2. AETIOLOGY

Modes of infection

As far as is known *Myco. leprae* does not multiply outside the human body, either *in vitro* or on animal inoculation. In open cases bacilli are shed from the nose and mouth and ulcerating sores, and in some cases from the unbroken skin surface. They are transmitted directly by contact or droplet infection, or indirectly through clothes, furniture and so on, and possibly also by insects. It is believed that bacilli enter the body principally through abrasions of the skin or nasal mucosa. It is not known whether infection takes place by way of the mouth or the gastro-intestinal tract. Many cases are known of leprosy acquired after accidental inoculation, but experimental inoculation in man has failed on several occasions to produce the disease (Rogers and Muir, 1946).

Susceptibility

There seems little doubt that a small minority of people are particularly susceptible, but whether or not this susceptibility is hereditary is not known. There is evidence that super-infection can break down natural resistance, and that young children are more disposed to infection than adults.

History absent

History of contact with infection is often absent, but this is not to be wondered at considering the long incubation period, averaging 3½ years but sometimes extending to 20 years, and the fact that many cases become infectious long before the disease is recognized. In most countries, though not in all, leprosy is commoner in adult males than females. Lowe (1934) considers that this may depend upon physiological as well as sociological differences between the sexes.

Sex

Low infectivity

Leprosy is of low infectivity compared with the sister disease tuberculosis; it tends to die out when a community becomes "leprosy conscious", or when the sanitation and the standard of living rise above a certain level.

3. PATHOLOGY

Appearance and staining of bacillus

Commonly known as the lepra bacillus, *Myco. leprae* is 1–8μ in length, and may be rod-like, diphtheroid or beaded in appearance. Our knowledge of it is limited since, as noted above, it does not multiply *in vitro* or on animal inoculation (McKinley, 1939).

It is Gram-positive and is acid-fast and alcohol-fast. In biopsy smears taken from a typical case and stained by the Ziehl-Neelsen method, the most characteristic appearance is the "globus", consisting of bunched bacilli held in a rounded mass of lightly stained material. This is contained in the so-called "lepra cell".

Organs involved

Leprosy affects the skin and subcutaneous tissue, the mucous membranes of the mouth and respiratory passages down to the larger bronchi, but not the lungs. The gastro-intestinal tract below the glottis is exempt. The liver is attacked but its functions are seldom noticeably impaired. Of the other internal organs the testis is most affected, its functions being destroyed sometimes. The kidneys are frequently involved in advanced leprosy as a consequence, not of bacillary invasion, but of toxaemia following secondary infection.

Nervous system

The central nervous system is exempt, but many of the peripheral nerves become infiltrated with bacilli, the infection reaching them through the blood-stream or, more frequently, by passing up the sensory nerves from skin lesions into the larger mixed nerves. Tissue reaction to these bacilli causes pressure upon nerve fibres and consequently sensory, trophic and vascular changes in the parts supplied by the nerves. This gives rise to polyneuritic changes in the extremities, with anaesthesia and trophic changes in the bones, joints and muscles of the hands and feet.

Eye

The anterior portion of the eye is affected, the infection spreading from the surrounding skin and invading the sclera, cornea and uveal tract. The optic nerve, retina and choroid are seldom, if ever, affected (Vase, 1946).

Histopathology of types

The type of disease and histopathological picture vary according to the degree of resistance which the subject is able to oppose to the infection. When resistance is low the bacilli multiply often to huge numbers and form "leproma" with "foamy" or lepra cells, surrounded by small, round cells. Hence this type is known as "lepromatous". When resistance is high the bacilli are few and difficult to find. Sections show epithelioid and giant cells with small cell infiltration, as in chronic lesions of tuberculosis. Hence this type is known as "tuberculoid". There is also an "uncharacteristic" type which, like other chronic inflammations, shows only a small round cell infiltration.

4. CLINICAL PICTURE

General signs

Leprosy, depending upon the resistance of the patient, may be severe and progressive, or slight and abortive. The disease generally begins with one or more circular patches of varying size, distinguished from the surrounding skin by one or more of the following characteristics: loss of pigment, erythema, thickening, loss of tactile, thermal or pain sensation, anhidrosis and loss of hair.

Lepromatous type

In the low-resistant "lepromatous" type the disease may be heralded by a flat, uncharacteristic lesion with ill-defined margin, slight reddening or

blanching and faint loss of feeling. This may disappear and be followed by multiple, symmetrically distributed flat macules, which gradually enlarge and



FIG 166.—Lepromatous type of leprosy, with discrete nodules.



FIG. 167.—Lepromatous type, with flat nodules on face and arms.



FIG. 168.—Tuberculoid type. View of the back, showing small discrete tubercles



FIG 169.—Tuberculoid type—*showing widely distributed lesions.*

coalesce effecting diffuse, more or less superficial, infiltration of wide areas of the skin. Later the infiltration deepens and the skin becomes thick and nodular (Figs. 166 and 167).

Tuberculoid type

In tuberculoid cases, which have developed resistance, the lesions (leprides) are more circumscribed, are limited by a raised margin and tend to heal at the centre (*Figs. 168 and 169*).

In this type the nerves are more affected, becoming thickened and tender and producing, as the larger mixed nerves of the limbs become involved, secondary sensory and trophic changes in the muscles, bones and joints of the hands and feet (Fig. 170). Leprides spreading over the face induce atrophy of the facial muscles, producing a mask-like face, sagging mouth and lagophthalmia.

Uncharacteristic type

While typical, fully developed lepromatous and tuberculoid cases are easy to differentiate, there is also an uncharacteristic type, intermediate in its



Fig. 170.—Tuberculoid type—showing trophic changes of hands and lips.



Fig. 171.—Uncharacteristic type, with serrated margin indicating rapid growth.

degree of resistance and with less definite clinical signs (Fig. 171). Leprosy of this type may be static, or may represent an initial transitional stage passing on into one of the more definite types. It may also mark an intermediate stage when one definite type is changing into the other, or when either of them is receding towards recovery.

Lepra reaction

Leprosy is ordinarily a chronic disease and the reaction of the tissues to the bacillus causes little or no distress. At times, however, reaction becomes suddenly acute, causing general febrile symptoms, with focal inflammation and even liquefaction and bursting of nodules and ulcer formation. This condition may be transient or may last for weeks or months. It is particularly distressing when the large nerves, eyes and nose are affected.

Eye

Leprosy of the eye is of particular importance, not only because of the frequency of blindness and impairment of vision, but because progressive or regressive tendencies of the disease can be conveniently studied in the transparent cornea, and through the cornea in the uveal tract. Keratitis may be punctate or pannus-like, or there may be deep nodule formation. The iris tends to be infiltrated, form posterior synechiae and become fixed, the pupil later becoming obstructed with exudate, resulting in total blindness.

In the tuberculoid type leprous infection is absent, but when lesions spread across the face they leave behind muscular atrophy and lagophthalmia, which in turn exposes the cornea to injury and sometimes [illegible]ation and ulceration. In neglected cases blin[illegible] infrequentl[illegible]

5. DIAGNOSIS

Diagnosis is based chiefly upon bacteriological and clinical examination.

Bacteriological examination

Myco. leprae are looked for in smears taken by scraping with the point of a knife the base of a small skin incision, or by scraping the nasal mucosa; they are also obtained from sputum or by lymph-node puncture. Smears are stained by the Ziehl-Neelsen method. Occasionally examination of skin sections or fibres teased from nerves is necessary to confirm diagnosis

Clinical signs

Clinically, inspection of the skin shows infiltration, widespread or in patches, characterized by colour changes, more or less thickening, keratosis or changes in the superficial skin markings, dryness of skin and loss or deformity of hair, nodulation in the lepromatous, and a mosaic of small tubercles in the tuberculoid, type.

Sensation (tactile, thermal and pain) is impaired to a greater or lesser degree, least in the lepromatous and most in the tuberculoid type of leprosy.

Testing sensory changes

Anaesthesia is best tested for by blindfolding the patient and asking him to put his finger on points touched with a feather or cotton-wool. Analgesia may be elicited by the "two-pin test", simultaneously pricking a normal and a suspected area of skin and noting which is the more painful.



FIG 172 —Tuberculoid type—showing thickening and nodulation of auricular nerve

Changes in nerves

Thickening and tenderness of the larger superficial nerves (especially the ulnar, radial, superficial peroneal and auricular) (Fig. 172) may be elicited by palpation, and of the smaller sensory nerves in relation to skin lesions by percussion; these manipulations also cause tingling in the region of distribution. Caseation and abscess formation occasionally occur in nerves, subcutaneous tissue and tendon sheaths.

Early diagnostic tests

In early doubtful lesions the histamine test is of value, the affected skin area failing to give the usual flare when a 1 in 1,000 solution of histamine is pricked into it. Similarly when 0 2 cubic centimetres of a 1 in 1,000 solution of pilocarpine is injected into affected skin it fails to sweat. Friction of the skin may make an early lesion stand out, as it does not flush like the surrounding normal skin.

Lepromin test

The lepromin test, unlike the Mantoux test in tuberculosis, is of little use in diagnosis, as it is positive in a large proportion of those persons never exposed to leprosy. But in frank cases it is of considerable value in estimating resistance to the disease and defining the type. Local reaction to the antigen is in direct proportion to the patient's resistance, and is generally strong in the tuberculoid, negative in the lepromatous and weak or negative in the uncharacteristic type (Dharmendra and Lowe, 1946).

6. DIFFERENTIAL DIAGNOSIS

Other acid-fast bacilli

Care should be taken to distinguish *Myco. leprae* from other acid-fast bacilli. Smears from the nose may contain semi-acid-fast saprophytes, and *Myco. leprae* in the sputum may be mistaken for *Myco. tuberculosis*.

Resembling lepromatous type

The lepromatous type of leprosy should be distinguished from dermal leishmaniasis, diffuse sebaceous adenoma, blastomycosis and neurofibroma by the presence of acid-fast bacilli.

Tuberculoid

Tuberculoid leprides and uncharacteristic macules are differentiated from psoriasis, tinea, seborrhoeal dermatitis, syphilis, yaws, dermal leishmaniasis, lupus vulgaris, tuberculides, lupus erythematosus, leucoderma, scars of injuries and various dietary deficiencies and food poisonings chiefly by the clinical appearance, sensory changes and thickening and tenderness of nerves. In these two types bacilli are not often found but should be looked for.

Polyneuritic

Secondary polyneuritic lesions of hands and feet must be distinguished from syringomyelia by their loss of tactile sensation and sweat function and by the thickening of nerves; also from cervical rib, Raynaud's syndrome, and neuritis of septic, beri-beri or diphtheritic origin by the history, the presence of other signs of leprosy, the thickening of nerves and the nature of the sensory and trophic changes.

7. PROGNOSIS

In a disease which, at its worst, breeds such dread, and yet in its milder forms may be transient and heal in a few months, an accurate prognosis is of great importance. Favourable indications are the tuberculoid type, few and limited lesions, a strongly positive lepromin test, and the possibility of improving and maintaining the patient's general health. There are indications that treatment with sulphones, at present under trial, considerably increases the chance of recovery from the severe lepromatous type.

8. GENERAL AND MEDICAL TREATMENT

General

As in tuberculosis, general treatment is of great importance. Removal of accompanying diseases, improved nutrition, occupational therapy, healthy climate and cheerful surroundings—none of these can be neglected if the best results are to be obtained.

Chaulmoogra oil

Chaulmoogra (hydnocarpus) oil and its derivatives have for long been and still are the standard treatment for leprosy. These are injected intradermally, intramuscularly and subcutaneously in doses of from 2 to 6 cubic centimetres (some recommend up to 10 cubic centimetres) once or twice a week. For the milder types of leprosy (tuberculoid and some of the uncharacteristic forms) intradermal chaulmoogra oil is still the best form of treatment.

Sulphones

Most promising results have been obtained recently with drugs of the sulphone group (promin, diasone, promizole, Sulphetrone) in the treatment of the severe lepromatous type. It is still too soon, however, to estimate the final value of these remedies (Muir, 1946 and 1947).

9. SURGICAL TREATMENT

Excision of early lesions

Excision of one or a few, small, definitely limited tuberculoid skin lesions may eradicate the disease, but equally good results can generally be more easily obtained by intradermal injections of chaulmoogra oil into the lesions.

6. DIFFERENTIAL DIAGNOSIS

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9 SURGICAL TREATMENT

Excision of early lesions

[illegible] may [illegible]

obtained by intradermal injections of chaul[illegible]

There is reason to hope that the use of sulphones in the earlier stages of leprosy will increasingly make operative treatment of the skin, feet, nose and eyes less necessary.

Sulphones

Aesthetic as well as ameliorative improvement can be gained by shaving off nodules and the outer layers of thickened skin and by trimming the ears; this is followed by the application of pure carbolic acid or some other caustic.

Shaving off nodules

Painting the skin with 1 in 3 trichloracetic acid aids resolution of lesions. Multiple skin puncture with thermocautery has been recommended for the same purpose.

Caustics and cautery

Splitting or removal of a thickened nerve capsule, or division of hard connective tissue binding down the ulnar behind the elbow, or the superficial peroneal at the neck of the fibula, may relieve pain and sometimes restore function. Abscesses of nerves, subcutaneous tissue or tendon sheaths, occurring in tuberculoid cases, may have to be opened. They contain pus or white liquefied caseous material, and generally acid-fast bacilli also, though not as a rule in large numbers.

Nerve operations

Loss of sensation, atrophy of the small muscles of the sole and consequent injury, often cause ulcers of the sole which develop secondary infection and, in the end, cause necrosis of phalanges, metatarsus and sometimes the tarsal bones. Often these trophic or "perforating" ulcers will heal with ordinary dressings, provided the affected part is kept off the ground by padding the sole, or by the use of crutches. Dead or decalcified bone should be removed without delay, otherwise septic absorption and immobilization of the patient have deleterious effects.

Bone operation

Partial or total metatarsectomy can be done through an incision on the dorsal or plantar surface. Anaesthesia of the foot generally makes an anaesthetic unnecessary. After removal of bone, and trimming, bleeding is controlled by packing the wound tight with a sulphonamide gauze and applying a few temporary stitches for 48 hours. The wound after healing should be allowed to consolidate for a few weeks before the foot is put on the ground.

Metatarsectomy

When the distal tarsi are diseased, Syme's amputation will often give good results.

Syme's amputation

Nodulation, ulceration, scar-contraction and obstruction of the nose often cause great distress. If dilation with bougies, cauterization and other milder treatments do not suffice, the obstruction must be removed by paring away the lepromatous lining, including the affected turbinate bones. A rubber tube is worn for 1 or 2 months till healing takes place.

Nose operations

Infected lacrimal sac is not uncommon in leprosy. It should be removed by the external route and an opening made into the nose with a small curette.

When active disease has ceased, nasal deformities may be improved by a plastic operation; an ivory plate should be inserted.

In the more severe forms of leprosy vision may be improved and blindness averted by timely action. The pupil should be dilated with atropine before posterior synechiae form and the pupil becomes fixed. Increased intra-ocular pressure may be prevented by iridectomy, and this operation may be more safely done if the tendency to reaction has been controlled by a previous course of one of the sulphones.

The eye

Acute lepra reaction in the eye can be relieved by injecting a 1 in 1,000 solution of trypan blue, so as to balloon up the bulbar conjunctiva.

Lagophthalmia In lagophthalmia, a sequela of the tuberculoid type of the disease, the eye should be protected by instilling liquid paraffin drops by day and bandaging on wool soaked in liquid paraffin by night. In some cases the eye may be protected by lateral tarsorrhaphy.

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LIGATURES AND SUTURES

By T. T. STAMM, F.R.C.S.
ORTHOPAEDIC SURGEON, GUY'S HOSPITAL, LONDON

1. INTRODUCTION

220] Ligature material is used for two main purposes:

(1) As ligatures to arrest bleeding.

(2) As sutures to approximate divided structures and obliterate dead space.

(1) Function

Importance of natural reparative reactions

In all cases permanent arrest of haemorrhage and repair of tissues depend upon the natural reparative reactions of the body. Ligature and suture materials merely hold structures in position while natural repair of the damage is brought about. These considerations determine both the features which it is desirable that such materials should possess, and the general principles upon which the technique of their use should depend.

(2) Technique

Avoidance of tension

With certain exceptions structures should never be sutured under tension. The tension of the suture will cause local arrest of the blood supply, and this

will inhibit the process of repair and may even cause local necrosis of the tissues, with subsequent "cutting out" of the sutures. For this reason a suture holder is one of the most dangerously convenient of instruments in the hands of the inexperienced or unimaginative surgeon. The surgeon must always have in mind the state of affairs which will obtain some days after the operation, and not merely the appearance of the part as he sews it up.

The only exceptions to the above rule are:

Conditions in which tension is justifiable

(a) In bone surgery, when strong sutures applied under tension may be necessary to hold fractures in position securely enough to resist the subsequent pull of muscles, for example, in fractures of the patella.

(b) In dealing with tissue loss after injuries, when temporary stay sutures, applied under some degree of tension, may sometimes be justifiable.

(c) *In abdominal surgery, when tension sutures may be inserted to resist* expulsive efforts, such as coughing. Even then the so-called tension sutures should not be tight.

(d) In re-making torn ligaments, when the new ligament may be temporarily reinforced with strong sutures drawn tight.

Fine sutures preferable

The suture material chosen should be the finest that is consistent with efficiency. The use of coarse sutures implies the necessity for tension and, except in the instances mentioned above, this nearly always means bad surgical technique.

Moreover, coarse sutures are more likely to interfere with the healing process, and to set up tissue reaction. When approximating tissues, many fine sutures are far more effective and less likely to lead to local areas of necrosis than are a few coarse sutures. Even when ligating main vessels several fine sutures spaced a little apart are more certain than is a single strong suture.

Cutting ligatures at the knot

Ligatures should always be cut at the knot, so as to leave the minimal amount of foreign material in the wound. If the knot is likely to slip sufficiently to take up any loose ends the ligature will be ineffective in any event. The only exception to this is in the case of catgut. As this material becomes moist in the wound the knots always slip to some extent; provided that the ligatures do not actually come undone this slipping is partly made up for by the swelling of the material when it becomes wet.

2. DESIRABLE FEATURES OF SUTURE AND LIGATURE MATERIALS

The following are qualities especially to be sought in choosing materials for sutures and ligatures.

(1) The materials must be easy to sterilize without loss of physical qualities

(2) Preferably, they should be absorbable, but if they are unabsorbable they should be chemically and physically inert in the tissues.

(3) They should possess good tensile strength, in relation to cross-section, combined with flexibility.

(4) The surface of the material should be so smooth that it causes the minimum of trauma when drawn through the tissues, yet knots must remain secure.

There are two main groups of materials, absorbable and unabsorbable.

3. ABSORBABLE MATERIALS

(1) Catgut

Although research is at present being conducted on the development of synthetic absorbable sutures, none of these materials has yet passed the experimental stage and catgut remains the only absorbable suture which is available for general use. For this reason alone, it is still more widely used than is any other material in spite of its many disadvantages.

Research on synthetic absorbable sutures

(a) Advantages

Apart from the fact that it is absorbed by the tissues of the body, catgut has no special merits compared with any of the other materials in general use.

(b) Disadvantages

(i) *Difficulty of sterilization.*—Catgut is manufactured from animal tissues, namely the elastic and muscular coats of the sheep's intestine. On this account the utmost care must be taken in its sterilization so as to make certain that all spores are killed, and this can be undertaken only in specially equipped laboratories.

(ii) *Variability.*—As a result of these manufacturing difficulties there are very considerable variations in the quality of the material, as regards both tensile strength and absorbability.

(iii) *Low tensile strength.*—Because of its low tensile strength as compared with that of all the unabsorbable materials, sutures of adequate strength tend to be clumsy and bulky.

Clumsiness and bulk

(iv) *Tissue reactions.*—It is the reactions provoked by the tissues which lead to the absorption of catgut, and these reactions are the inevitable result of the use of any absorbable suture.

They will result in its absorption only if conditions are favourable. In a wound that becomes septic the suture may be surrounded by a purulent exudate. In such an event, absorption will not take place and the suture will act as a foreign body in the same manner as would an unabsorbable suture. Although a risk of sepsis is usually regarded as an indication for the use of catgut, and although catgut is preferable to unabsorbable materials in such cases, it must be remembered that it cannot be used with impunity.

(v) *Knots do not hold well.*—Catgut becomes very slippery when it is wet and knots are very liable to slip. This feature is less marked in those varieties that have been hardened by immersion in a solution of chromic acid. The hardening process, however, does render it less pliable and sympathetic to use.

Chromicized catgut

(c) Special uses

Catgut is especially suitable in the following circumstances.

(i) In septic or potentially septic wounds since it is likely to be absorbed and thus will not remain as a foreign body.

(ii) As a continuous suture for the repair of circular tubes or moving planes, when continuous non-absorbable sutures would remain, so limiting the circumference of the tube and the extensibility of the plane.

Continuous suture for repair of circular tubes

(iii) For skin sutures when the removal would be painful, or is dreaded; for example, in circumcision, in scrotal incisions and, sometimes, for children.

(iv) In those situations in which the presence of an unabsorbable suture might lead to mechanical or chemical irritation of the tissues, for example,

will inhibit the process of repair and may even cause local necrosis of the tissues, with subsequent "cutting out" of the sutures. For this reason a suture holder is one of the most dangerously convenient of instruments in the hands of the inexperienced or unimaginative surgeon. The surgeon must always have in mind the state of affairs which will obtain some days after the operation, and not merely the appearance of the part as he sews it up.

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There are two main groups of materials, absorbable and unabsorbable.

(iii) They are also contra-indicated in cases in which even a slight amount of tissue reaction is undesirable, such as in nerve suture.

(iv) Since their structure is similar to that of a wick they are liable to lead infection from one part of a wound to another if used as a continuous suture, or from the skin to deeper structures if used for skin suture.

(v) Since their surface is not smooth, they cannot be drawn through delicate tissues without producing a certain amount of trauma.

(c) Special uses

Although widely employed in the past, their use in modern surgery should be restricted almost entirely to the ligation of blood-vessels. The security of the knot makes for the peace of mind of the surgeon when ligating large vessels, and for multiple ligations they have the advantage of simplicity and speed in use; for these purposes, therefore, their disadvantages are of little significance provided that the wound is clean.

Ligation of blood-vessels

(d) Sterilization and storage

The main supplies of multifilaments are usually kept unsterile, sterilization being carried out by boiling for ten minutes when the material is required for use. They can be kept sterile in any of the usual antiseptic solutions, such as a 1 in 20 solution of carbolic acid.

(3) Impregnated multifilaments

An attempt has been made to overcome some of the disadvantages of the unabsorbable materials already mentioned by impregnating them with wax or some form of plastic material. This overcomes their tendency to act as wicks and to harbour micro-organisms. They are therefore much to be preferred to untreated multifilaments, though they retain the other disadvantages of this type of material.

Wax or plastic material

Special uses

There are certain special uses to which impregnated multifilaments are put. These are:

(i) For the ligation of arteries.

(ii) For arterial and cardiac surgery when the ease and rapidity with which they can be handled, and their security, are important features.

(iii) In abdominal surgery for those cases in which the merits of an absorbable suture are outweighed by the insecurity and unreliability of catgut.

(4) Monofilaments: general

This group comprises natural salmon gut, the synthetic materials rayon and nylon, and metal filaments.

(a) Advantages

Monofilaments, in general, possess the following advantages:

(i) They cannot harbour organisms in the substance of the material.

(ii) There is no capillary action along the thread.

(iii) They are inert in the tissues.

(iv) Owing to these features they are less likely than are multifilaments to cause trouble in an infected wound and they do not prevent its healing.

in joint and other cavities, where it might cause friction, or in the biliary and urinary tracts, where it might cause the formation of calculi.

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Catgut should always be purchased already sterilized and packed in sealed glass ampoules. The ampoules can be kept ready for use in a 1 in 20 solution of carbolic acid, or in any other suitable antiseptic solution. If such ampoules are not available it is safer to use an unabsorbable material rather than to attempt to sterilize the catgut locally, unless special facilities are available.

Carbolic acid solution

4. UNABSORBABLE MATERIALS

(1) General considerations

These materials all have certain advantages over catgut. These advantages are that they:

(a) Can be sterilized by heat.
(b) Have far greater tensile strength.
(c) Excite little or no tissue reaction.
(d) Can be tied securely and cut at the knot.

The individual varieties of such materials can be considered most conveniently under four headings.

(2) Multifilaments

This group comprises silk, linen and cotton thread. Although each of these materials has its almost fanatical supporters their general features can be considered together.

(a) Advantages

The following are the advantages of multifilaments:

(i) They are the easiest and most convenient materials to handle and use, and are the only ligature materials that can be conveniently used on a reel.

(ii) In proportion to their diameter, their tensile strength is very great.

(iii) Knots made with them show little or no tendency to slip. The loose ends, therefore, can safely be cut at the knot, thus leaving the minimum of foreign material in the body.

(b) Disadvantages

Disadvantages which should be considered in assessing the value of this material are:

(i) The interstices between the fibres provide shelter for bacteria and protect them from the defences of the body. This will delay the healing of an infected wound and may lead to the formation of a sinus.

Delayed healing of infected wound

(ii) Multifilaments are not entirely inert in the tissues and do provoke a minor degree of foreign-body reaction. In a perfectly healthy, clean wound this may be without significance, but if any organisms are present it may lead to the formation of a residual abscess, either at the time, or even years later. Although this risk is a small one, it means that, whilst it is justifiable to use such materials for the ligation of vessels, or for small interrupted sutures, they should not be used for continuous sutures as this entails leaving too extensive a foreign body in the wound.

Minor degree of foreign-body reaction

Contra-indicated for continuous sutures

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Minor degree of foreign-body reaction

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disadvantages that it is radio-opaque, and may be a hindrance if subsequent exploration of the same area is necessary.

Special uses.—It is used by some surgeons in preference to nylon for nerve suture and for plastic work. The heavier gauges are especially useful for "removable" sutures in the repair of tendons, and also in orthopaedic work for the fixation of bones.

(d) Tantalum wire

The characteristics of this material, which has been introduced comparatively recently, are similar to those of stainless-steel wire, but it is rather more pliable. Its trials have not yet been sufficiently extensive to permit a final assessment of its merits.

TABLE

TENSILE STRENGTHS OF SURGICAL THREADS

Material		Mean diameter (in millimetres)	Breaking load (in kilograms)	Breaking stress (in kilograms per square centimetre)
Plaited silk	No 5	0 38	3·678	5·20
	No. 4	0 36	3·150	3·18
	No. 3	0 32	2 730	3·33
	No. 2	0 22	1 428	3 90
	No 1	0 16	1·168	5·53
Chinese twist	No. 5	0 63	6 978	2 23
	No. 4	0 45	4·178	2·58
	No 3	0 33	1·978	2·31
	No. 2	0 33	2·628	3·07
Linen thread	No. 25	0 36	2·828	2·78
	No. 35	0 31	2 068	2·78
	No 40	0 27	1·878	3 18
	No 60	0 23	1 878	4 56
	No 90	0 17	1·078	4 75
	No 100	0 17	0 538	2·40
Deknatel	Very fine	0 10	0 378	4·81
	Fine	0 16	1·028	5·11
	Medium	0 24	1 378	2·95
	Strong	0 23	1·478	3·56
Nylon	Fine	0 24	1·148	2·62
	Medium	0 30	1·628	2 30
	Strong	0 37	2 378	2·22
Waxed thread	Medium	0 19	1·118	3·94
	Thick	0 22	1 078	2·84
Silkworm gut	Fine	0 28	1·050	1·69
	Medium	0 38	1 520	1·35
	Strong	0 40	1 478	1·18

The above figures were worked out in the Physics Department of Guy's Hospital. They are the actual figures of samples taken at random from the surgical theatre and indicate the variations in breaking strains which occur even with the same type of material

5. SUMMARY

Nylon monofilament is probably the best material available for most general surgical purposes, and especially for skin sutures and for plastic and nerve

(b) Disadvantages

On the other hand, the following disadvantages should be borne in mind:

(i) They are less easy to handle than are multifilament threads.

(ii) Knots do not hold so well in some cases, especially when the coarser grades are used.

(iii) They have less tensile strength than have multifilaments with the same diameter.

(c) Special uses

The following are special uses to which monofilaments may be put:

(i) They are the most suitable materials for skin suture.

(ii) For the repair of soft parts when it is necessary to use a considerable quantity of suture material as in the repair of hernias and tendons.

(iii) As an alternative to catgut when it is necessary to bury a continuous suture.

(iv) In orthopaedic surgery for the fixation of fractures, and so forth.

(d) Sterilization and storage

Boiling undesirable for synthetic material

All these substances can be sterilized by boiling, but in the case of the synthetic materials repeated boiling is undesirable. After sterilization they can be stored in a suitable antiseptic solution, such as 1 in 20 carbolic acid, with the exception of nylon which is destroyed by carbolic acid, and must be kept in some other antiseptic, such as a 2 per cent solution of chlorocresol.

Metal filaments, when required for immediate use, are usually sterilized by boiling with the surgical instruments.

(5) Monofilaments: individual features

(a) Salmon gut

Disadvantages

Made from the bodies of silkworms, this material is stronger than rayon ar nylon, but it is obtainable only in short lengths, is variable in quality a tends to become brittle with repeated sterilization and storage. It has be... largely replaced by nylon and stainless-steel wire.

(b) Rayon and nylon

These materials have less tensile strength than other monofilaments, but they are available in any length of constant diameter. They are more pliable than salmon gut, but knots do not hold so well except in the case of the finer gauges.

Nerve suture and plastic work

Special uses.—Nylon is the most pliable monofilament and the least traumatic of all suture materials. It is also completely inert in the tissues. It is, therefore, an admirable material for nerve suture and for plastic work, for in such cases only the finest gauges need be used, and with them there is little danger of the knots slipping. It is also useful for the repair of small tendons and for all skin sutures.

(c) Stainless-steel wire

Kinking

This material is completely inert in the tissues and can be sterilized repeatedly by heat without any loss of its physical qualities. It has great tensile strength, but it is difficult to handle and has a tendency to kink. Kinking must be carefully avoided, as it always causes a weak spot in the wire. It has the further

LIMBS—ABSENCE OF

By R. D. LANGDALE KELHAM, O.B.E., M.R.C.S., L.R.C.P.
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1. GENERAL

221] It is only in recent years that the practice of supplying artificial limbs to children has been more generally accepted.

Formerly, a child born without a hand or whose hand had been amputated for any reason, was not given an artificial arm or hand, with the result that when adult age was reached he had no desire for one, having become so accustomed to "making do" with one hand. If a foot was amputated a peg leg was supplied.

Psychological effect

The lack of an artificial arm or the fitting of a peg leg as a permanent measure is liable to have a bad psychological effect upon the parents and, at a later date, upon the child; the absence of a hand or presence of a peg leg is a constant reminder to the parents that their child is not as other children are, and consequently they treat him as a cripple. The consciousness of abnormality, and the treatment accorded to him by others, influences the child adversely at a later date when he would be associating with normal children in play and at school He becomes acutely conscious of his physical differences and limitations and, depending upon his personality, becomes resentful or develops a sense of mental segregation full of unspecified longings, and, whilst desirous of joining in games, is afraid to do so for fear of exhibiting inferiority.



Fig. 173.—A boy with an amputation above the elbow, writing with an artificial hand.

Early fitting in children

Artificial limbs not only can but should be supplied to children at the earliest possible age; this is, for arms, immediately prior to going to the first infant school, and for legs, as soon as the child would normally walk. The artificial legs are miniature replicas of those supplied to adults. For infants they can be made in wood, with a rigid ankle and metatarsal joint, and for those of 8 years of age and over, in metal with all joints fully articulated.

surgery. When it is necessary to use a strong suture, stainless-steel wire is preferable as the knots hold better, and it gives more rigid fixation in orthopaedic work. Waxed threads are widely used for the ligation of arteries and for arterial and cardiac surgery owing to their security and the facility of their use. Catgut should be used only when it is essential, or highly desirable, that the material employed should be absorbed, its main uses being for the suture of the deeper layers of a wound or in cases in which infection is feared.

[References to other titles are given under Ligatures and Sutures, in the Index Volume.]

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2. ARTIFICIAL ARMS

Artificial arms for infants are in the first instance cosmetic arms with ...



FIG 174.—A girl with a forearm amputation, using a typing appliance.

children can engage in various handicrafts and hobbies, both at school and at home. (Fig. 174.)

Congenital stumps

Frequently, congenital upper and forearm stumps are met with in which a rudimentary thumb or finger is growing from the surface of the stump. It is rarely necessary to remove these outgrowths, many of which do not interfere with the artificial limb. Many cases arise of congenital deformity of the hand in which only a thumb and perhaps one rudimentary digit remain. In some cases these rudiments are very useful, but the parents desire an artificial hand in order to conceal the deformity. In such a case a cosmetic hand or appliance should be made which will enhance the value of the natural grip and retain the sensitivity of the remaining digits.





FIG. 175. FIG. 176

A boy aged 2 years, fitted with a pair of artificial legs. The right is a "kneeling" leg. When the right tibia had grown sufficiently, a normal artificial leg for a below-knee amputation was fitted.

3. AMPUTATIONS OF THE LOWER EXTREMITY

For amputations of the lower extremity, the principles of weight bearing and of lengths and types of stumps remain exactly as for adults, but it is not

always possible to provide the best length of stump for a child in the first instance. When surgical conditions permit, it is best to disarticulate through the knee and fit a limb for this condition until such time as the femur has grown sufficiently to permit of an amputation at the site of election.

Below-knee amputation

For amputations below the knee a stump of 5½ inches is desirable but, when not obtainable in the first instance, it has been possible to fit a kneeling leg until such time as the tibia has grown sufficiently to permit the fitting of a leg for below-knee amputation. (Figs. 175 and 176.) The child illustrated was so fitted at 2 years of age, but has since been given a pair of below-knee legs. Daily quadriceps exercises are essential to avoid flexion deformity at the knee being induced by the wearing of a kneeling leg.

Daily exercises





FIG. 177 FIG 178.

A pair of artificial limbs (right, "kneeling" type, left, "platform" type) supplied for a child with a bilateral congenital deformity. When the left tibia has grown sufficiently, the foot will be amputated and an artificial leg for a below-knee amputation fitted.

It is not always necessary that children or adults should continue to use unsightly and revealing surgical boots and appliances, nor, for some of the cases, is amputation necessary before a more normal-appearing artificial limb can be supplied. In a number of congenital malformations, and particularly in old cases of

Congenital malformations

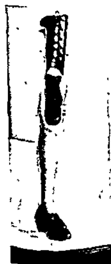






FIG 179. FIG 180 FIG 181.

Types of "platform limbs" supplied to adults who had suffered from anterior poliomyelitis in childhood.

infantile paralysis, there is a foot or rudimentary foot, upon which weight can be taken, together with a grossly shortened tibia or femur, or both. For such cases a "platform limb" can often be supplied in which weight is taken

"Platform limb"



Fig. 182.—Shortening of right leg and deformity of foot.

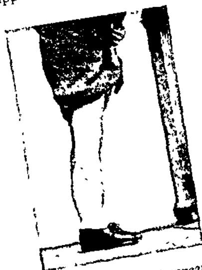


Fig. 183.—Abnormal appearance of leg when wearing appliance.



Fig. 184.—Showing how an artificial limb can be fitted to conceal deformity, without amputation (Side view.)



Fig. 185.—Front view

upon the deformed foot around which the artificial leg of normal length is built, with an artificial foot and an ankle joint.

Figs. 177 and 178 show artificial limbs for a bilateral congenital deformity in which the right tibia is too short for anything but a "kneeling" leg at present, and a limb has been provided for the left short tibia with the deformed foot. When this leg has grown sufficiently, the foot will be amputated and the usual type of below-knee leg supplied, but in the meantime the child gets about very well with these legs.

LIPOID METABOLISM AND LIPOID GRANULOMA

By A. H. T. ROBB-SMITH, M.D.
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1. DEFINITION

The lipoidoses

222.] The term "lipoid" is used to include the triglyceride esters of the fatty acids, cholesterol and cholesterol esters and the phospholipoids.

The disturbances of lipoid metabolism to be considered here are particularly the "lipoidoses" in which there is a deposition of lipoid in the tissues of the body, commonly in reticular cells or histiocytes, which may or may not be associated with the presence of abnormal amounts of lipoids in the blood.

A lipoid granuloma is a circumscribed proliferation of connective tissue cells associated with a deposition of lipoid.

2. NORMAL LIPOID METABOLISM

It would not be appropriate to attempt a detailed discussion of normal lipoid metabolism (Mitchell, 1946) but it may be said that in an ideal diet 20–30 per cent of the calories are in the form of fat. This is absorbed from the small intestine, in part being transformed to phospholipoid, and is transported in the blood stream to the fat depots—the adipose tissue and the liver—whence it can be mobilized and modified for energy metabolism or as building stones of the cytoskeleton, such as mitochondria and cell membranes. Apart from the formation of lipoid from dietary fat, the body lipoids can be synthesized both from carbohydrate and from protein.

3. CLASSIFICATION OF DISTURBANCES OF LIPOID METABOLISM

(1) Effects of a deficiency of dietary fat—starvation and emaciation.
(2) Effects of an excess of dietary fat—obesity.
(3) Disturbances in the absorption of fat—steatorrhoea.

Intestinal lipodystrophy (mesenteric chyladenectasis) is an uncommon disorder occurring exclusively in males who, after suffering from mild arthritis for years, begin to lose weight in spite of a good appetite, develop steatorrhoea and may present with a palpable mass of enlarged lymph nodes in the upper abdomen which may result in exploration for a tumour. Severe cases are invariably fatal and at post-mortem examination there is a diffuse thickening of the serosa of the intestine with considerable mesenteric lymph node enlargement. Microscopical examination reveals dilatation of the lymphatics, which are filled with fat, and lymph-nodal fibrosis (Clemmesen, 1945).

4. DISTURBANCE IN THE DISTRIBUTION OF THE FAT DEPOTS

These are commonly associated with endocrinal dysfunction, though there is the rare condition of *progressive lipodystrophy*, occurring exclusively in young fema.es, in which there is a complete absence of subcutaneous fat of the face, neck and trunk, but a normal or often excessive amount of subcutaneous fat from the pelvis downwards. Although there is an appearance of extreme emaciation, the patients show no signs of ill health or weakness.

5. LIPOIDOSES

In this group of disorders, as a result of disturbances of lipoid metabolism, there is a deposition of abnormal amounts of lipoid in the cells of the body, either restricted to reticular cells or histiocytes (cells of the reticulo-endothelial system) or affecting other cells as well. They may be divided into three main groups: (1) the primary lipoidoses in which there is an inborn error of lipoid metabolism; there is some doubt as to whether the deposition of the lipoid in the cells should be regarded as a reflection of a general disturbance of intermediary lipoid metabolism in which there is either an excess production or a diminished breakdown of the lipoid and the cellular deposition is a

Main groups of lipoidoses

secondary change or, as appears more likely, that there is a primary disorder of intracellular metabolism in the reticular cells, (2) the secondary lipoidoses in which there is an abnormal deposition of lipoid consequent upon a general metabolic disorder such as diabetes mellitus, and (3) the secondary lipoidoses associated with progressive reticular hyperplasia, such as the Hand-Schuller-Christian syndrome and eosinophilic granuloma (Thannhauser, 1940).

(1) Primary lipoidoses

Three main forms are recognized, depending upon the type of lipoid laid down in the cells.

(a) Phosphatide lipoidosis

Two varieties are recognized, both exceedingly rare and of little surgical import.

Niemann-Pick's disease is a familial disorder occurring almost exclusively in female infants of Jewish ancestry. There is gross enlargement of the liver and spleen with emaciation and a brownish-grey pigmentation of the skin; there is commonly mental retardation, and death from intercurrent infection occurs usually before the second year.

Amaurotic familial idiocy is also familial and is characterized by blindness—associated with a cherry-red spot in the region of the macula—and idiocy associated with progressive muscular weakness. Symptoms may be manifested in early infancy or may be delayed until adolescence.

(b) Cerebroside lipoidoses

Gaucher's disease is uncommon but, as surgery gives symptomatic relief, it deserves a little attention. The disease commonly manifests itself in childhood, but there is an infantile form coming on in the first few months of life with hepatosplenomegaly, mental retardation and muscular rigidity, death occurring in a few months; the infantile form is familial.

In its more usual form, the children or young adults present with a gross splenomegaly but with comparatively good health; there is often a brownish pigmentation of the skin, and conjunctival pingueculae. Radiographic examination will often reveal a broadening of the lower end of the femur so that the condylar prominences are less marked (Erlenmeyer flask deformity) and occasionally there is a widespread bony involvement with the appearance of cystic spaces in the long bones, kyphosis due to vertebral collapse and spontaneous fractures. Blood examination will reveal a mild hypochromic anaemia, leucopenia and occasionally thrombocytopenia, sometimes with haemorrhagic manifestations; there may be a hyperochlesteraemia. The diagnosis can be made by the finding of the typical Gaucher cells (reticular cells 20–40 μ in diameter with a small nucleus and a bluish skeined cytoplasm) in sternal or splenic puncture preparation. The differential diagnosis is that of a gross chronic splenomegaly in a child or young adult.

Splenectomy renders the patients much more comfortable and possib
prolongs life; apart from the size of the spleen, the operation is seldom di
cult although the possibility of complicat orrhage must be conside
in cases in which there is a thrombocyt y of the patients surv
for 20 or more years after diagnosis and paratively normal l
and many of the splenectomized patie through pregna

without difficulty. It is not inherited and though there may be a familial tendency it is not very marked.

The average weight of the spleen is 3,000 grammes. Its shape is maintained and the cut surface is a reddish snuff colour, often with areas of infarction Microscopical examination reveals a diffuse medullary replacement by the Gaucher's cells, while there is commonly siderosis of the littoral cells in the venous sinuses. Gaucher's cells are also to be found in the liver, lymph nodes and bone marrow but it is unusual to find other organs involved.

(c) *Cholesterol lipoidosis (xanthomatosis)*

Two varieties of primary xanthomatosis are recognized, depending upon whether or not there is a raised blood-cholesterol level.

Hypercholesteraemic xanthomatosis

The hypercholesteraemic form of essential xanthomatosis may occur at any age and may be familial; although skin lesions are the commonest manifestations, these are often associated with changes in the internal organs. The skin lesions range from xanthelasmas of the eyelids, occurring as yellowish-orange plaques and situated usually at the medial border of the lids, to tuberose xanthoma which are found as multiple nodules on the extensor surfaces, usually at friction or flexure points; the nodules may appear and disappear. Sometimes this is associated with xanthomatous involvement of the tendons and may be mistaken for gout. In association with the skin lesions there may be a xanthomatosis of the heart and vessels and such patients may die suddenly from cardiac failure. In women there is an association between skin xanthomas and a xanthomatous infiltration of the biliary ducts inducing intermittent jaundice and biliary cirrhosis; there may be splenomegaly as well. In all these cases there is invariably a considerably degree of hypercholesteraemia and the serum may be milky. Histological examination of the tissues reveals collections of xanthoma cells (histiocytes or reticular cells filled with lipoid) and a variable degree of fibrosis.

The treatment is essentially dietetic and, on the whole, disappointing. Splenectomy has not been shown to be of value, and the surgical importance of this group of conditions is chiefly from the point of view of differential diagnosis.

Normocholesteraemic xanthomatosis

In the normocholesteraemic type of xanthomatosis, the skin lesions are of the disseminate type, usually reddish-brown in colour and present in the axillae, around the neck and ante-cubital fossa and may ultimately be scattered over the body. Occasionally there may be infiltration of the pituitary fossa with the occurrence of diabetes insipidus, although this is more commonly associated with the lipoid granulomas of the Schüller-Christian type (*see* p. 430).

(2) Secondary lipoidoses associated with general metabolic disorders

In certain general metabolic diseases there may be a considerable increase in the circulating neutral fats, rather than in cholesterol, and this is often associated with xanthomas and hepatosplenomegaly of varying degree. The conditions in which this may occur are familial idiopathic hyperlipaemia, diabetes mellitus, chronic pancreatitis, von Gierke's disease (glycogen storage disease), nephritis of the nephrotic type (chronic nephrosis, Type II nephritis of Ellis) and others.

(3) Secondary lipoidosis associated with progressive reticular hyperplasia

There is a group of disorders, occurring in children and young adults, of varying degree of chronicity which have common morphological features in that there are proliferations of fibrous tissue and reticulum cells, which have been known as the Letterer-Siwe syndrome, Hand-Schüller-Christian syndrome, eosinophilic granuloma of bone and lipogranuloma, and are recognized now to have a common morphogenesis although their aetiology is quite unknown.

It will be most convenient to describe the three syndromes although there are numerous examples of intermediate types.

(*a*) *Letterer-Siwe syndrome* (*acute reticulosis of infancy*)

This condition occurs in young children, seldom over the age of 4 years, and is characterized by an irregular pyrexia associated with lymphadenopathy and hepatosplenomegaly. In addition there are destructive lesions in the bones—frequently in the skull but the long bones may be involved as well—and a papular purpuric eruption on the trunk, becoming more marked in the terminal stages; there is often a chronic otitis media, ulceration of the skin and other signs of secondary infection. There may be respiratory signs associated with a honeycombed appearance on radiography. There is an increasing hypochromic anaemia, leucopenia and thrombocytopenia. The condition is invariably fatal, with a usual duration of a few months, but there may be remissions so that the course of the disease may be prolonged to a ar or more. Microscopical examination reveals a widespread proliferation f reticulum cells and histiocytes in foci in the affected organs, with a variable legree of fibrosis; there is commonly a moderate lipoid infiltration of the histiocytes with the formation of xanthomatous foci. The treatment is entirely symptomatic and no cures have been reported.

(*b*) *Hand-Schuller-Christian syndrome* (*lipogranulomatosis of bone*)

This is a chronic disorder, commonly occurring in childhood though symptoms may be delayed until adult life, and having as its basic symptomatology, calvarial defects ("geographical skull"), exophthalmos and diabetes insipidus; in addition, there may be defects in other bones, skin eruptions and pulmonary involvement characterized by diffuse fibrosis. In addition to diabetes insipidus, there may be signs of dyspituitarism with genital hypoplasia, stunting of growth and obesity. The diagnosis may be confirmed by biopsy of an affected bone when, in the chronic lesions, there will be fibrous tissue in which are lipoid-containing histiocytes, with reticulum cells and a varying proportion of haemic cells; however, the early lesions show no xanthomatosis but merely a fibro-reticular proliferation (Fig. 186). This fibro-reticular tissue is present extradurally, in the bones and the retro-orbital space, and analogous changes are found in the lungs; involvement of the lymph nodes, liver or spleen is unusual.

X-ray treatment has been found to be effective in the bony lesions and, by accelerating the fibrosis, it may diminish the endocrinal symptoms, but since the disease is commonly chronic with remissions it is difficult to judge the full value of such treatment.



Fi[illegible]



FIG. 187.—Eosinophil granuloma (170)

(c) *Eosinophilic granuloma of bone (solitary granuloma of bone)*

This condition occurs in children or young adults who present with localized pain and tenderness of bone; any bone may be affected, with the possible exception of those of the hands and feet though the most frequent sites are the skull bones, ribs, humerus or femur. There may be some swelling at the site of tenderness, but no redness or heat. Radiographic examination reveals a circumscribed area of translucency with very little reaction or enlargement of the bone, though there may be vertebral collapse or pathological fracture. In some cases there may be a slight leucocytosis or an absolute eosinophilia without leucocytosis. There are usually no systemic symptoms or evidence of involvement of internal organs, but radiography may reveal lesions in bones other than that which has given rise to symptoms. On biopsy the area o affected bone will be found to contain soft brownish granulation tissue whic may appear necrotic. Microscopical examination reveals sheets of reticulu cells and histiocytes interspersed with numerous eosinophil leucocytes. Th may be a moderate number of osteoclast type giant cells, some of which c tain iron pigment, and some fibrosis, but xanthoma cells are scanty (Fig. 1 The prognosis is good, curettage usually being sufficient to induce healing, and in some cases x-ray irradiation of the order of 1,500 r in divided doses over a period of 6 days has resulted in healing though this is commonly slower than with surgical intervention, and there is evidence to suggest that spontaneous resolution may take place.

The aetiology of this group of disorders is quite unknown. It is clear that the primary lesion is the proliferation of reticulum cells with a propensity to fibre formation and that the xanthomatosis is a secondary phenomenon. The eosinophilia is present to a variable degree in all three types although it is most marked in the solitary granuloma. In the acute reticulosis of infancy and in the eosinophilic granuloma, both the clinical and the pathological features are suggestive of an infective aetiology, but all bacteriological investigations have as yet been unsuccessful (Jaffe and Lichtenstein, 1944).

6. SECONDARY LIPOIDOSIS ASSOCIATED WITH TRAUMA AND INFLAMMATORY CONDITIONS

In this group of conditions, as a result of a local alteration of the fatty tissue, the neutral fats are split to form fatty acids and glycerol or abnormal fats are introduced, either of which may induce a slowly progressive sclerosis with panniculitis.

(1) Traumatic oleogranulomas

As the result of the introduction of paraffin or oils into the subcutaneous tissues, a fibrogranulomatous lesion results which tends to be progressive and which extends through the tissues although there is commonly a latent period of months or years before the nodular masses develop. The acute lesions are present in the "grease-gun hand" in which, as the result of the injection of oil into the tissue spaces of the fingers, ischaemia and gangrene may result. The more chronic lesions are the paraffinomas following cosmetic operations, the injection of camphor in liquid paraffin or of phenol in oil for the treatment of haemorrhoids, and a few cases in which accidental occupational injection of oil has induced late oleogranulomatous lesions; localized

oleogranulomas have been described as occurring on the scalp, resulting from an excessive use of brilliantine.

The late effects of oleogranulomas are always serious, and it is essential, whenever practicable, to excise the affected area as widely as possible soon after the accident, for in the later stages excision of extensive nodular tumorous masses may be well-nigh impossible (Moore, 1946).

Fat necrosis (*see* Fat Necrosis, Vol. 4, p. 84).

(2) Panniculitis

Panniculitis represents a more diffuse and chronic fat necrosis in which a traumatic aetiology is not apparent.

(3) Sclerema neonatorum

This is characterized by widespread masses of hard fat in the subcutaneous tissues in the new-born infant, which on microscopical examination are seen to consist of sclerosis with areas of fat necrosis. The origin of the condition is obscure but obstetrical trauma is almost certainly not the main factor (Zeek and Madden, 1946).

(4) Relapsing febrile nodular non-suppurative panniculitis (Weber-Christian disease)

This condition usually occurs in adult females. It is manifested by the occurrence of raised, tender, reddish nodules, ranging from a few millimetres to several centimetres in diameter, usually on the thighs and abdomen and is associated with an irregular pyrexia. After a while the nodules regress to leave depressed scars; the disease is chronic with remissions and exacerbations. Histological examination reveals an acute inflammation of the fatty tissue with subsequent scarring and the few post-mortem examinations have shown that the panniculitis is not restricted to subcutaneous fat. The aetiology of the condition is unknown and there is no specific treatment (Ungar, 1946, Baumgartner and Riva, 1945)

(5) Rothmann-Makai type of panniculitis

In this condition there are no systemic symptoms, but there is the sudden appearance of small, soft, slightly tender, symmetrically disposed, subcutaneous nodules which may persist for days or weeks and then slowly disappear without leaving any scars (Baumgartner and Riva, 1945).

(6) Necrobiosis lipoidica diabeticorium

Diabetic patients occasionally develop, chiefly on the legs, circumscribed red, hard, peripherally growing, raised papules which tend to show a central depression with scleroderma-like consistency, sometimes there is ulceration at the centre. Histological examination reveals a necrosis of the dermis with a diffuse fatty infiltration (Klaber, 1934).

Insulin lipodystrophy

These lesions are quite distinct from insulin lipodystrophy and insulin lipomas, which are probably a peculiar form of traumatic fat necrosis (Rowe and Garrison, 1933).

7. XANTHOMATOUS CHANGE IN ASSOCIATION WITH NEOPLASTIC HISTIOCYTIC PROLIFERATIONS

There are a number of conditions, characterized by local proliferation of histiocytes, which may be regarded either as tumours or as progressive

hyperplasias in which on occasion the histiocytes contain much lipoid material, so that xanthomatous areas are present. These can be considered quite briefly since they are only variants of conditions which are dealt with elsewhere.

(1) Giant-cell tumour of bone

On occasion the giant-cell tumour of bone has a yellowish-white appearance rather than the characteristic brownish-red colour and on histological examination it is found that in addition to the multinucleate giant cells in the cellular fibrous stroma there are numerous lipoid-containing histiocytes.

(2) Villo-nodular synovitis, bursitis and synovitis

In these conditions there is a yellowish-brown nodular thickening of the tendons and synovial membrane which on microscopical examination shows proliferation of iron-containing and fat-containing histiocytes (Jaffe, Lichtenstein and Sutro, 1941).

(3) Histiocytoma

These are small, circumscribed, cutaneous tumours of a greyish tinge, sometimes showing hyperkeratosis on their surface, often appearing to infiltrate the epidermis and occasionally showing a depressed centre. They occur most frequently on the extremities and on excision are seen to be yellowish-brown in colour. Microscopically they consist of closely set fibrocytes and histiocytes containing lipoid and iron particles. They are benign but are apt to be mistaken for melanomas. (Arnold and Tilden, 1943).

8. LIPOMATOSIS, LIPOMAS AND LIPOSARCOMAS

Circumscribed adiposity

Apart from diffuse increase of fatty tissue resulting from dietary, metabolic or endocrinal disturbances, there are certain conditions of circumscribed adiposity which merge imperceptibly into the circumscribed fatty tumours or lipomas.

(1) Symmetrical diffuse lipomatosis (Madelung's neck)

This occurs almost exclusively in middle-aged males, usually of alcoholic tendencies, who in other respects are in good health. If the fatty masses are excised, they do not recur though lipomatosis may appear elsewhere.

(2) Adiposis dolorosa (Dercum's disease)

Adiposis dolorosa, on the other hand, is limited to females. The fat deposits may be diffuse or nodular, symmetrical or irregular and may occur in any site although the face, hands and feet usually escape. There is usually asthenia, with signs of vasomotor instability and psychic abnormalities in addition to the neuritic pains (Wells, 1940).

Circumscribed lipomas may often be extremely tender. Copeman and Ackerman (1944) have shown that in many cases of "fibrositis", the trigger-points or tender rheumatic nodules are due to herniation of fat through the enveloping fibrous membrane and that excision of these herniations will relieve the symptoms.

(3) Renal lipomatosis

Renal lipomatosis is the most significant form of visceral lipomatosis and is invariably a sequel to renal lithiasis (Dukes, 1938).

(4) Lipomas

Lipomas are probably the commonest benign tumours and may reach an enormous size. They can occur in any site, and in certain situations such as the nervous system (Krainer, 1935), thorax (Heuer, 1933), gastro-intestinal tract (Botsford and Newton, 1941; Rumold, 1941), or kidney (Robertson and Hand, 1941) they may be of especial surgical significance. Lipomas in the neck may be of brownish colour and associated with lymphangiomatous tissue; they are regarded as being derived from the foetal fat glands (Inglis, 1927). *Site*

Multiple lipomas may be familial and may sometimes show a nerve distribution or are closely related to nerve fibres; other mixed types of lipomas—fibro-lipomas and lipo-angioma—may occur (Adair, Pack and Farrier, 1932; Tedeschi, 1946).

Large lipomas may undergo degeneration, in particular calcification, and although malignant change may occur in a benign lipoma, it is excessively rare.

(5) Liposarcomas

Liposarcomas are an uncommon form of malignant connective-tissue tumour. Their recognition is important because with adequate surgical excision in the early stage a cure may be achieved, but too often their malignant character is not recognized until surgical excision is impossible. They are most commonly reported from the retroperitoneal or mesenteric region, but, as Stout (1944) has shown, the lower limbs are probably the most frequent site of occurrence. *Sites of occurrence*

The gross appearance is variable but commonly they are encapsulated, lobulated, semi-translucent tumours with areas of mucoid character, interspersed with white or yellowish nodules, and occasionally are cystic or haemorrhagic. It is not uncommon for isolated nodules of tumour tissue to lie adjacent to, but separate from, the main tumour mass. In the initial stages the tumours do not show marked invasive characters but there may be attritive changes in the organs in the vicinity.

The histological characters are very variable. In places the tumour cells may resemble foetal or granular fat cells, elsewhere they may be spindle-form cells with isolated fatty globules usually in a mucoid stroma and highly cellular forms with giant cells and active mitoses in which the lipomatous origin may be difficult to recognize. In the past there have been attempts to distinguish between myxo-lipomas and myxo-liposarcomas, but in a single tumour every type of cellular pattern may be seen.

The speed of growth of tumours is extremely variable but on the whole they grow slowly, although frequently the spread of growth is accelerated with repeated recurrences. The chief character of liposarcomas is their tendency towards local or zonal recurrence, largely due to the apparent encapsulation of the tumour with a consequent tendency to a close local excision. Distant metastases are less common except in the highly cellular forms, in which there may be secondary deposits in lungs or liver.

The treatment of liposarcomas is radical surgical excision and in the event of a tumour having been locally excised and being proved on histological examination to be a liposarcoma, it is essential that a further and more radical excision—even amputation if necessary—be performed if recurrence is to be avoided. Radiotherapy has not been shown to be of any value. *Treatment*

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LIVER—CIRRHOSIS

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1. INTRODUCTION

223.] There is no direct surgical treatment for portal cirrhosis of the liver *per se*, and operative intervention is never a primary method of election in this condition. Nevertheless, for many years surgeons have exercised ingenuity in devising means of short-circuiting the portal blood flow in order to diminish or obviate the secondary effects, such as ascites and spontaneous haemorrhage, which result from progressive fibrosis around the vessels in the liver.

2. METHODS OF TREATMENT

One of the earliest suggestions was to by-pass the liver by making a direct communication between the portal vein and the vena cava, as had been done experimentally in animals as long ago as 1877 by the physiologist Eck, this direct communication is frequently referred to as "Eck's fistula". When first applied to the human subject by Vidal and others in cases of cirrhosis, the results were very unsatisfactory, the patients usually dying in cholaemia. This was supposed to be because the portal blood, charged with the products of intestinal digestion, passed into the systemic circulation without purification in the liver.

Development of vascular adhesions

It was the observation, in the post-mortem room, of the development of numerous vascular adhesions between the viscera and the parietes in patients who had died of cirrhosis without ascites, or in whom that latter condition had disappeared, which led *Drummond and Morison in 1895 to devise the* method of omentopexy with the object of imitating Nature in short-circuiting the portal circulation by the development of collateral channels between the portal and systemic venous systems (Drummond and Morison, 1896). Proof that this can be done has been amply furnished by the after-progress of many

Omentopexy

cases, and by post-mortem observations. In the early stages of the development of this form of therapy the operation was much abused; it was carried out in all kinds of cases of ascites of undetermined pathology and with poor results. When limited to cases of alcoholic cirrhosis without such complications as cardiac or renal disease, and to cases in which the patients have withstood several tappings without the onset of cholaemia or metabolic liver breakdown, the results have been most encouraging. The infrequency with which the operation is carried out at the present time is not a measure of its lack of success, but is due to the rapidly diminishing incidence of alcoholic cirrhosis which was so common 30 or 40 years ago.

Dietetic methods

The close survey of the biochemistry of the blood in cirrhosis has now led to a better understanding of the metabolic upset resulting from this condition, and it has been found that by attention to a proper liver dietary regimen and by other measures, the condition of the patients can be very much improved, provided always that the causative factor ceases to operate. In most cases, however, the complications of spontaneous haemorrhages and ascites are only partly controlled by such therapy and for their relief, or as a prophylactic measure, the making of a direct porto-caval anastomosis or shunt has been revived. The results are now being closely studied.

Porto-caval anastomosis

If this direct method is employed, a short circuit is made either by uniting the portal vein itself to the vena cava or by uniting one of the large contributory branches such as the splenic branch to the caval system *via* one of its tributaries, usually the renal vein. Such a junction is capable of allowing a large volume of blood to by-pass the obstruction Incidentally, the removal of the spleen is estimated to eliminate about 40 per cent of the blood volume passing through the liver. Technically, porto-caval anastomosis is a difficult procedure which requires much judgement, experience in vascular surgery and great technical skill. Failure may result when the shunt is not large enough or when the union becomes occluded by thrombosis. Great improvement, however, has sometimes followed such intervention and the problem is to decide when it should be employed (Blakemore, 1947).

Wasting ascites and --rhage

The conditions that bring cirrhosis of the liver into the surgical orbit are uncontrollable wasting ascites and spontaneous haemorrhages. Though wasting ascites almost invariably goes on to a fatal termination, its progress is usually slow and is to be measured in months. Haemorrhages, on the other hand, are always alarming and often result in the immediate death of the patient or else frequently lead up to that event when there is the usual recurrence of bleeding.

3. PRELIMINARY INVESTIGATIONS

All cases of cirrhosis of whatever origin should be closely studied in co-operation with a physician, for it may be possible by biochemical studies, directed to liver function and to the condition of the blood, to determine what part may be played in the production of symptoms by depressed and disordered liver function, and what part may be attributed to hypertension in the portal venous system. A study of urinary output and renal function may give valuable information. Liver biopsy is also said to have been useful. When the porto-caval shunt is under consideration, it is essential to have the information provided by excretion urograms since nephrectomy may have

to be performed when spleno-renal anastomosis is considered necessary. It is only during recent years that the importance of oesophageal varices as a source of serious spontaneous haemorrhages has been realized, and much valuable information may be gleaned from the study of oesophagrams and the use of the oesophagoscope. Great improvement may follow medical management; the main measures are to institute a well-regulated liver dietetic regimen which largely consists in reducing the fat intake to a minimum, increasing the protein intake, and making up the calorific value by carbohydrate. At the same time, the vitamin intake is increased, especially that of vitamins A, D and K, and of vitamin B complex. The regulation of fluids and the proper use of diuretics and cardiac stimulants are also of importance.

Oesophageal varices as source of haemorrhage

Under such a regimen many cases improve considerably; ascites may absorb and spontaneous haemorrhages diminish in frequency and severity. If such improvement does not occur or falls short of sufficient relief, or if there is anxiety because of repeated haemorrhage or recurrent ascites then surgical measures should be considered. The two plans of attack are (a) omentopexy and (b) porto-caval anastomosis or shunt.

4. OMENTOPEXY

This method is also known as epiplopexy, epiplorrhaphy or the Talma-Morison or Drummond-Morison operation. The object of the operation is to form new vascular communications between the systemic or parietal and the portal or visceral venous systems.

The operation was devised for the relief of ascites and with no other object in view. Its possible effect on the other main result of portal hypertension—spontaneous haemorrhage—has not been particularly studied after omentopexy, but it can be stated that in the patients who have survived successful operation for some time, spontaneous haemorrhage has not been a prominent event. It might be expected that the establishment of an effective collateral circulation, primarily directed to the cure of ascites, would equally have relieved the serious hypertension with varicosity and engorgement in the vessels of the lower oesophagus and cardia.

(1) Indications for operation

Drummond and Morison advised that their operation should be reserved for ascites resulting from alcoholic cirrhosis, and they stipulated that the patients selected should be free from jaundice and cardiac or renal disease, and should have withstood several tappings. When these rather rigid indications have been observed the results have been very satisfactory. In ascites due to syphilis or polyserositis the results have been unsatisfactory. In other types of ascites, often of undetermined origin, the results have also been unfavourable, but there is no reason why the operation should not be tried when other methods have failed. Success is not to be expected, whatever the primary cause, unless the ascites is mechanical and not merely toxic; success is also unlikely if the patient is already suffering from cholaemic symptoms or is rapidly deteriorating. When the ascites is a symptom of Banti's disease, splenectomy combined with omentopexy has been followed by survival for several years without recurrence of ascites, until death has resulted from progressive liver deterioration.

Alcoholic cirrhosis

Ascites of undetermined origin

Banti's disease

(2) Immediate preparation

These patients are often bad risks, and a week or two should be spent in close co-operation with a physician, who can superintend the dietetic and general management of the embarrassed liver. For some days before operation patients should be kept in bed so that they become accustomed to lying on their backs. Bowel action should be free, and efforts made to overcome distension. The abdomen should be re-tapped about 4 days before the intended operation.

(3) Technique

It is difficult to perform the complete operation properly without a general anaesthetic, although some surgeons have insisted on the superiority of local anaesthesia. The original operation comprised three definite steps:

(1) An attempt to promote the formation of vascular adhesions between the liver and spleen and the parieties.

(2) Fixation of the spread-out omentum to the peritoneal aspect of the abdominal wall for the same purpose.

(3) Drainage of the peritoneal cavity and strapping of the abdomen so that the viscera may be kept in contact with the parietal peritoneum while the anticipated adhesions are forming.

Incision

The incision should be a median one from the ensiform cartilage to within an inch or so of the umbilicus. The peritoneum is opened to the left of the midline in order to avoid the falciform and round ligaments of the liver, with the contained enlarged and engorged veins met with in cirrhosis. For the same reason the incision should stop short of the umbilicus with a view to avoiding injury to the caput Medusae. On opening the peritoneal cavity the first step is to inspect or palpate the liver, in order to verify the diagnosis. Great care must be taken not to tear or break down any existing adhesions.

Drainage

A small independent opening is then made above the pubes, just large enough to introduce a Keith's glass drainage tube of ½-inch diameter (Fig. 188) which is to be introduced to the bottom of the recto-vesical or Douglas's pouch, where it remains. Any fluid which does not escape by the tube is removed with the suction apparatus or is soaked up in mops. The escape of the fluid and the inspection of the viscera are facilitated by the use of the reversed Trendelenburg position.

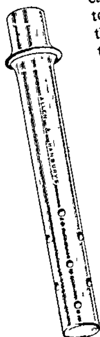


FIG 188.—Keith's drainagetube. (*Allen & Hanburys Ltd.*)

rmation of 'hesions

As soon as the main part of the fluid has been removed, the surfaces of the liver and spleen and the adjoining parietes are vigorously scrubbed with gauze with the deliberate object of injuring the endothelial covering in order to encourage the formation of adhesions. The spread-out omentum is then fixed to the posterior peritoneal surface of the abdominal wall over an area about the size of the palms of both hands, that is, for about 3½–4 inches on either side of the midline incision and about the same distance below the umbilicus (Fig. 189). This may be done by everting

the abdominal wall, which will probably be sufficiently lax after the escape of the fluid, and attaching the omentum to the parietes at several points by a series of interrupted sutures or by a couple of rows of continuous suture on either side (Fig. 190). Another convenient method is by means of silkworm-gut sutures passed through the omentum and through the whole thickness of the abdominal wall and tied outside over rolls of gauze or pieces of drainage tubing. About 3 or 4 such sutures on either side of the midline will probably suffice. The omentum is also caught and sutured to the parietal peritoneum during the closure of the incision, which is to be done with care, as incisional hernia has often been an unpleasant sequel. As a last step, the abdomen is carefully strapped from above downwards and firmly bandaged over a light dressing, in order to keep the parietes and viscera in contact. The tube is left *in situ* and is carefully surrounded and covered by gauze soaked in a reliable antiseptic, such as 1 in 1,000 perchloride of mercury, and an independent dressing.

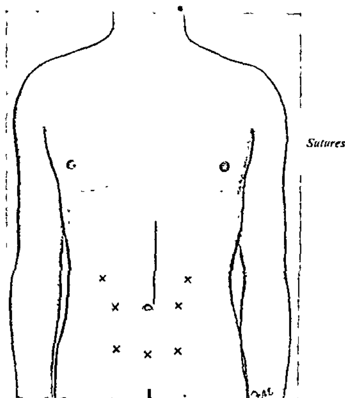


FIG 189—Situation of abdominal incision and points of attachment of omentum to parietes

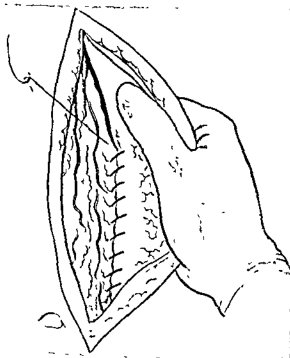


FIG. 190—Suture of omentum to peritoneal aspect of anterior abdominal wall The suture is repeated on the opposite side (*B*) *courtesy of W. B. Saunders & Co. Ltd*)

5. POST-OPERATIVE CARE

From the first, the patient must be kept propped up in bed to encourage fluid to gravitate to the pelvis. The most important consideration is to guard against infection and this is done by the intelligent use of reliable antiseptics. In these days, penicillin will probably be administered prophylactically. The gauze about the tube will absorb some fluid and should be changed when soaked. The fluid which accumulates in the pelvis must be withdrawn, with a syringe or the suction apparatus, at first every hour, and subsequently at increasing intervals. In 3 or 4 days it diminishes so much that only half an ounce or so can be withdrawn by the syringe every 4 hours. When this stage arrives, it is sufficient to apply gauze over the mouth of the tube and to change it as often as it becomes soiled. The glass tube should be rotated twice daily to prevent omentum becoming entangled in the holes and to release any masses of lymph which may block them. A rubber tube may be substituted for the glass one on about the fourth day; one that will just easily occupy the lumen of the Keith's tube is slipped into the latter, which is then withdrawn. After the substitution of the tubes, the patient may be moved in bed more freely. All drainage can be dispensed with on about the tenth day.

Withdrawal of fluid

Cholaemia

During the post-operative period the greatest danger is from cholaemia, which may come on about the second to the fourth day. This should be guarded against by continuous study of the blood chemistry, by watching the urinary output, by securing early movement of the bowels and by abundance of fluids by the mouth and the intravenous use of 5 per cent glucose solution or, if response is tardy, by blood transfusion. As soon as the risk of cholaemia is over, the amount of fluid administered should be diminished as much as possible. There is some risk of pulmonary complications, and the patient should, therefore, be encouraged to take deep breaths and should be assisted to cough systematically. Unless there has been leakage of fluid through the median incision, this need not be dressed until the twenty-first day, when the silkworm omental fixation sutures and the through-and-through sutures in the incision may be removed. At the end of a month the patient may leave bed, but the abdomen should be kept firmly bandaged for some weeks. Even when drainage by tube has been successful, there is usually some re-accumulation of fluid; this may be absorbed, though it may be necessary to tap the abdomen once or twice. Progress towards recovery may be rather slow, possibly occupying a period of some months rather than weeks but, even so, may ultimately be complete.

Pulmonary complications

Re-accumulation of fluid

It is essential that the patient should abstain entirely from all alcoholic liquors and that any other causative factor should be removed if the operation is to be permanently successful. Several of the patients have become milk-drinkers or have been very fond of sucking sweets. For some time after the operation there may be a tendency to hepatic insufficiency, as shown by periods of lethargy with slight icterus. This must be guarded against by the appropriate dietetic and other management. Persisting oesophageal varices may be the cause of haematemesis.

Hepatic insufficiency

6. RESULTS OF OPERATION

The immediate mortality in suitable selected cases used to be about 15 per cent, but with better understanding of the management of liver insufficiency

this should be much less. Symptomatic cures have been obtained in about 40 per cent of cases. Many patients have greatly improved in health after operation and have lived and worked in comfort for years. Two patients who were rapidly going downhill in spite of careful treatment and repeated tapping are known by the writer to have been alive and perfectly well 15 and 34 years, respectively, after operation (Turner, 1940). Recurrence of the ascites may follow a return to alcoholic habits, but secondary cure may again take place after abstention and suitable treatment. The results have been best when the operation has been carried out comparatively early in the disease.

Recurrence of ascites

(1) Sequelae

Some of the male patients have complained very much of inguinal hernia, which has usually been present before the operation but has been overshadowed by the more troublesome ascites. The fact that such patients are anxious for radical cure is perhaps evidence of the increasing self-respect which results from their restored health. Incisional hernia has occurred in several of the cases and may be very troublesome.

Incisional hernia

Operations for such hernias have proved hazardous because there is a tendency to cholaemia for some time after the operation. The abdominal wall is also unusually vascular, as the result of venous engorgement. Any subsequent surgical intervention should be deferred for 6 months.

After recovery, patients are, of course, exposed to the usual hazards which follow any laparotomy, but there has not been any special incidence of intestinal obstruction by bands or adhesions. Should such a condition arise it must be dealt with surgically, without delay. On account of the tendency to liver insufficiency in the early post-operative months, it would be wise at that stage to use local or spinal anaesthesia.

(2) Modifications

The most important modification is the omission of the drainage tube. Of course, this greatly simplifies the after-treatment, but in these circumstances the fluid may be expected to re-accumulate until adhesions become vascularized, and the abdomen may have to be tapped several times in the first few weeks or months after the operation. If tapping has to be resorted to frequently and at short intervals before operation, a period of drainage is wise. Some operators prefer to fix the liver edge to the parietes by suture rather than to rely on the possible formation of adhesions. Occasionally the omentum is shrivelled and lies rolled up close along the colon, leaving no "apron" for fixation to the abdominal wall. In these circumstances the transverse colon should be stitched to the parietal peritoneum. Schiassi raised a flap in the abdominal wall and fixed the omentum and spleen into a space made between the posterior surface of the muscles and the peritoneum, whereas Mayo introduced a portion of the omentum into a pocket made by separating the posterior sheath of the rectus from its muscle. Narath's method aimed at combining subcutaneous drainage with the formation of new vascular channels, and for this purpose he introduced the omentum into the subcutaneous tissue of the abdominal wall, but this plan is certain to be followed by troublesome and potentially serious hernia.

Omission of drainage tube

7. SECONDARY OMENTOPEXY

Secondary omentopexy may be required when there has been a previous laparotomy, and it is not unlikely that it may have to be carried out in some cases in which attempts to bring relief by porto-caval anastomosis have failed to provide complete relief.

In these circumstances, it is imperative that the surgeon should interfere as little as possible with adhesions which have already formed and which may contain venous communicating channels. For this reason, the abdomen should be opened by an incision between the midline and the centre of the rectus, or in any event through a fresh site from the previous laparotomy. Curiosity as to the conditions resulting from any previous operation should be curbed for the same reason. If there are no omental adhesions the ordinary technique can be carried out, but it may be that existing vascular bands over the liver or spleen will make it unwise to interfere with either viscus. Efforts should then be concentrated on securing adhesion of the omentum over as wide an area of parietal peritoneum as possible. If that membrane is whitened and thickened, it may be deeply scratched or even incised here and there down to the underlying muscle, with the object of bringing the omentum freely into contact with vascular tissue.

Incision

If the abdominal wall is unusually vascular, a rapid method of dealing with venous bleeding is to apply a continuous suture of catgut along its margins, including the whole thickness except the skin. Just before closing the abdomen this suture may be relaxed piecemeal and individual vessels caught and tied but if that is likely to take too long or to be very troublesome, the edges of the abdominal wall with the continuous catgut *in situ* may be brought together with interrupted sutures, including its whole thickness, introduced about ⅜ inch apart. Such sutures ought to be left *in situ* for not less than a fortnight, at the end of which time union will probably be quite sound.

Control of venous haemorrhage

8. PORTO-CAVAL ANASTOMOSIS

Technique

The first stage is an exploratory laparotomy in the upper half of the abdomen for the following purposes:

(a) To confirm the diagnosis.

(b) To demonstrate the precise conditions of the visceral veins.

(c) By manometric readings to determine the intravenous tension in the portal system.

The normal pressure in the portal vein is about 100 millimetres of water but it may rise as high as 500 millimetres. If the intervention is for haemorrhage rather than ascites and the portal pressure is very high, it is essential to have a sufficiently free shunt, or by-pass, so that a large vein like the portal itself must be anastomosed directly to the vena cava or a large radical like the splenic joined to a capacious systemic branch such as the left renal.

It has sometimes been considered that the removal of the spleen has in itself some beneficial effect on the metabolism of the liver disease. Its removal in Banti's disease is an essential part of the management of that condition. In establishing a porto-caval shunt it may be necessary to remove the spleen and left kidney to enable a direct end-to-end anastomosis to be made between

the splenic vein and the left renal vein though, by anastomosing the end of the splenic vein to the upper side of the left renal vein, the kidney may be spared. The actual union of the vessels is by direct suture when the lumina are large enough or, otherwise, over a special vitallium cannula which remains *in situ* (Fig. 191).

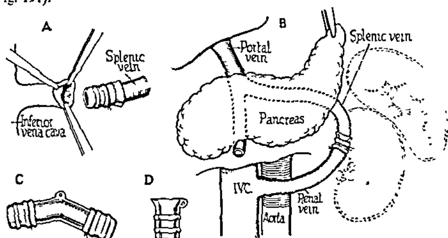


Fig. 191 —Diagrammatic representation of porto-caval anastomosis after splenectomy and nephrectomy. A: The method of union over a vitallium cannula. B The disposition of the vessels after anastomosis. C: Cannula lined with segment of vein to be used in bridging gap where the vessels cannot be directly united. D: Vitallium cannula (*B) courtesy of The Year Book Publishers Inc., Illinois.*)

The technical details of the actual anastomosis will be found in the section on Arteries (Vol. 1, p. 361) and are also described by Blakemore and Lord (1945) and by Whipple (1945).

These operations are tedious and exacting and may occupy 2 or 3 hours. Extreme care and gentleness are necessary to avoid injury to the vessel wall, which might precipitate thrombosis For the same reason, heparinization is required for some 24 hours after the intervention Secondary venous thrombosis is one of the sequelae to be feared Porto-caval anastomosis is still in the experimental stage but the results obtained in skilled hands justify further trials. The method has the advantage that technical failure or limited success does not rule out a subsequent trial of omentopexy.

Secondary venous thrombosis

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LUMBAR PUNCTURE

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1. INTRODUCTION

224.] The introduction of puncture of the spinal theca in the lumbar region by Quincke in 1893 led to immediate advances in the diagnosis of diseases of the nervous system. During the subsequent 50 years the procedure has not only increased its diagnostic scope but it has also become a valuable therapeutic adjunct.

2. TECHNIQUE

(1) Equipment

The special equipment required consists of a lumbar-puncture needle and a manometer. Several types of needle are available; the essential requirements are that the pattern should have a short bevel and a well-fitting stilette and that its diameter be not too great—that is, an external diameter of 1·2 millimetres. The Greenfield type of needle, in which there is incorporated a three-way tap, is useful in facilitating accurate manometry by minimizing the loss of cerebrospinal fluid. The manometer generally used consists of a length of glass tubing graduated in millimetres from 0 to at least 300, with an internal diameter not greater than 2–3 millimetres, so that the total volume of fluid required to fill the lumbar-puncture needle, the manometer and the short length of rubber tubing between them is not greater than 3–5 cubic centimetres.

Greenfield needle

Aseptic precautions

Puncture must be performed with every aseptic precaution. A wide area of the skin of the lower back should be cleansed with ether or treated with an antiseptic solution. Ideally the operator should wear sterile rubber gloves. When gloves are not available, the needle should be handled as little as possible—even after thorough preparation of the hands—and it should be held in a small sterile towel while the puncture is being made. The sterility of all the instruments and solutions used must be ensured. This implies that the lumbar-puncture needle and the manometer must be sterilized, either by boiling or in the electric oven.

(2) Position of patient

Usually the procedure will be performed with the patient in the lateral position, the lumbar spine being fully flexed and the line through the external

occipital protuberance and spinous processes horizontal. Only in such a position will accurate manometry be possible. In certain circumstances, for example when difficulty is experienced in entering the theca with the patient in a horizontal position, or when encephalography is intended, the procedure is carried out with the patient sitting up and the spine flexed as fully as possible.

Encephalography

(3) Method

With the patient in one of the positions described, an intradermal weal of local anaesthetic is raised over the interspinous interval at which the puncture is to be performed. This will usually be between the third and fourth or the fourth and fifth lumbar spines, the former being the interspace above and the latter that below a line joining the highest parts of the iliac crests. The lumbar-puncture needle is now inserted through the anaesthetic area, usually in the midline; in the presence of very large spinous processes it is inserted a short distance to one side of the midline. It is passed anteriorly and very slightly proximally, and at a depth of 5–6 centimetres in an adult a sensation of diminished resistance will be experienced as the point penetrates the ligamentum flavum. The stilette is then withdrawn and, if the dura and the arachnoid have been penetrated, fluid will drip from the needle. If it does not, the point must be slowly advanced until cerebrospinal fluid is obtained. The needle should not be forcibly passed into the spinal canal until its anterior wall is struck, since this is likely to lead to contamination of the cerebrospinal-fluid specimen with blood, and may cause severe leg pain if one of the spinal nerve roots is penetrated. The manometer is then connected and, with the patient relaxed and breathing quietly, the cerebrospinal-fluid pressure and its variations with cardiac and respiratory activity are noted. The increase in pressure with a forced expiration is also noted; a variation of less than 10 millimetres indicates that the point of the needle is not properly placed within the theca or that a nerve root obstructs its orifice. In such circumstances the needle must be readjusted before an attempt is made to determine the effects on the pressure of compression of each jugular vein independently and then of both together. This is the Queckenstedt response.

Variations in pressure

(4) Difficulties encountered

In the restless patient general anaesthesia may be required. When the lumbar spine is rigid and especially in the case of obese patients it may sometimes be difficult to enter the interlaminar interval. The sitting position mentioned above may then prove useful. On most occasions when cerebrospinal fluid is not obtained at lumbar puncture it may be assumed that the theca has not been entered. True dry punctures can occur, however, when the whole lumbar theca is filled with neoplasm or when the subarachnoid space at the level of the puncture is largely obliterated by an extradural intraspinal mass.

Obesity

Dry punctures

3. COMPLICATIONS

(1) Headache

The common complication is headache, to which vomiting may be added. It is often initiated by sitting up and is relieved by lying down; the cause is believed to be due to the continued escape of cerebrospinal fluid through the

puncture wound made by the needle in the spinal membranes. This low-pressure headache may at times be prevented by keeping the patient flat in bed for 24 hours after the puncture. Raising the foot of the bed and forcing fluids by the mouth are also useful therapeutic methods.

(2) Infection of meninges

Infection of the meninges after lumbar puncture is an extremely serious complication and one which is more common than is generally realized. It is probable that evidence of meningeal irritation after puncture is frequently due to this cause even when the symptoms disappear without treatment and no organism is cultivated from the cerebrospinal fluid. Numerous cases of infection of the meninges with Gram-negative bacilli, frequently with fatal results, have now been placed on record and there must be many more which have not been recorded. The risk of infection is increased when the puncture is used not merely to obtain a sample of cerebrospinal fluid but to inject some such fluid as a solution of penicillin or a spinal anaesthetic agent into the theca.

Infection with Gram-negative bacilli

(3) Injuries

The possibility of injury to spinal or intraspinal structures at the operation merits consideration. It is thought that injury to an intervertebral disc with subsequent degeneration and local symptoms has sometimes followed lumbar puncture. Although the piercing of an intrathecal or extrathecal nerve may occasion acute discomfort at the time, no permanent harm should result. However, if the puncture is carried out above the level of the second lumbar spine, injury to the spinal cord with permanent sequelae may be produced.

Injury to spinal cord

4. INDICATIONS

These fall into two groups: (i) diagnostic and (ii) therapeutic.

(i) *Diagnostic lumbar puncture.*—In diagnosis the procedure is of value in estimating intracranial pressure and in determining the presence of either partial or complete subarachnoid block. Furthermore, cerebrospinal-fluid examination may be of great diagnostic value. Finally, the injection of a radio-lucent or of a radio-opaque contrast medium, for example, oxygen or Lipiodol, through a lumbar-puncture needle is made in pneumo-encephalography and in myelography.

(ii) *Therapeutic methods.*—The therapeutic indications are several. The lowering of intracranial pressure which follows the procedure, and the removal of blood and pus from the meninges, may be helpful in cases of head injury or of meningitis. The injection of drugs, in particular penicillin, by this route is of the greatest value in the treatment of infections within the dural theca. Anaesthetic solutions are injected by this route in the production of spinal anaesthesia. It is suggested that in certain other conditions, for example, in status epilepticus, lumbar puncture may have a beneficial effect.

Injection of drugs

5. CONTRA-INDICATIONS

Infective lesions

Lumbar puncture is contra-indicated in the presence of infective lesions in the region of the puncture wound, whether such lesions are cutaneous, bony or

intraspinal, because of the danger of introducing infection into the sub-arachnoid space.

Raised intracranial pressure

The other important contra-indication is raised intracranial pressure. Although at times the procedure may be indicated, in the presence of such hypertension there is a real danger that when intracranial pressure is high sudden coma and death may follow the puncture. Therefore when all the evidence points to the presence of an intracranial tumour, and intracranial pressure is judged to be considerably raised, lumbar puncture should not be performed. In such circumstances it is dangerous and it gives little or no information of value.

[References to other titles are given under Lumbar Puncture, in the Index Volume. The subject is also dealt with under the heading of Cerebrospinal Fluid in the *British Encyclopaedia of Medical Practice* (1937), Vol. 3, p. 52.]

LUNG—TUMOURS

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INTRODUCTION

225.] Tumours of the lung may be either benign or malignant, malignant tumours forming about 90 per cent of the total. Carcinoma of the lung is the commonest. Adenoma of the bronchus is the commonest of the benign tumours and these two deserve the closest consideration.

PART I
CARCINOMA OF THE LUNG

Origin

All carcinomas of the lung arise in the bronchial epithelium, as was shown by Barnard (1926). The basal layer of the bronchial epithelium not only reproduces the columnar ciliated and goblet cells of normal epithelium, but by

metaplasia can also reproduce stratified squamous epithelium. This totipotency of the basal layer probably explains the pleomorphic character of these growths.

Physiological upset

Normal pulmonary physiology may be upset by encroachment on the alveolar area, either by the massive size of the growth, or by the production of collapse as a result of bronchial occlusion. A relatively small growth, for some reason as yet obscure, may produce severe dyspnoea even in the presence of the minimal degree of alveolar involvement; it was attributed by Tudor Edwards (1946) to tracheal fixation, but this cannot satisfactorily explain its disappearance after pneumonectomy.

1. PATHOLOGY

The growths can be grouped into two categories:

Morbid anatomy

(1) The main bronchus type of tumour, in which the mass is mainly endobronchial and usually affects the main, lobar or one of the segmental branches

(2) The circumscribed tumours which arise from the smaller grade bronchi form discrete tumours in the lung which have sharply defined margins and predominantly infiltrate towards the periphery of the lung either on the mediastinal or diaphragmatic surfaces or on the chest wall.

Very rarely, tumours at the apex of the lung invade the structures forming the thoracic inlet, giving rise to the syndrome described by Pancoast, which classically consists of erosion of the ribs and involvement of the brachial plexus and the cervical sympathetic. This was originally thought by Pancoast to be due to a specific type of tumour, but it has been shown to be due simply to the anatomical site of the tumour and may result not only from primary lung tumours but also from secondary deposits in the lung in this position.

The main bronchus type produces its symptoms by ulceration of the tumour and by producing occlusion of its bronchus, which leads to collapse with or without infection behind the block. When there is no infection behind the growth, a simple atelectasis of the area occurs; there is little dilatation of the bronchi though they may be full of mucus.

When infection is present, there may be extensive ulceration of the lung tissue behind the growth leading to lung abscess, or the bronchial tree may be very dilated with associated pneumonitis, with or without abscess formation.

Tumours of the circumscribed group are often silent, and when they give rise to symptoms do so by involvement of other structures such as the chest wall, or as a result of metastasis, and only later by ulceration into a larger bronchus. The circumscribed tumours, especially when of the squamous-celled type, are very prone to undergo central necrosis, the necrotic content being coughed up when the tumour has ulcerated into a bronchus; this not infrequently gives rise to an incorrect diagnosis of lung abscess. One other form of rupture into the bronchus gives rise to a projection of the growth, often of considerable length, along the bronchus. Bronchoscopically, it may be confused with a pedunculated tumour of the bronchus.

Histology

Histologically, the commonest type is a squamous-celled carcinoma, secondly the oat-celled type, and lastly the adeno-columnar-celled carcinoma.

The oat-celled tumours are the most *primitive, although* not uncommonly the squamous-celled group are of the anaplastic type. The adenocarcinoma *is the most fully* differentiated, although highly keratinizing squamous-celled



FIG. 192.—Section shows squamous-celled carcinoma of differentiated type with adjacent bronchial cartilage.



FIG. 193.—Section shows squamous-celled carcinoma with central degeneration of cells from a case showing well-marked central necrosis.



FIG. 194.—Section shows typical oat-celled carcinoma. Note variation in shape of cells from small round cells to the classical oat cells.

growths are met with. Often representative *areas of all these groups* can be seen in the same tumour. (Figs. 192–195.)

Dissemination

The tumours disseminate by blood-stream and lymphatic spread. It is generally agreed that blood-borne metastasis occurs most commonly with the circumscribed tumours, but diametrically opposed views are held as to lymphatic spread. Tuttle and Womack (1934) maintain that metastasis to parahilar and inferior tracheal lymph glands is earlier in the circumscribed than *in the* main bronchus *group*. Rabin and Neuhof (1934) maintain the converse. The writer's experience fully confirms the latter viewpoint, and the *probable explanation* is the presence of the intrapulmonary lymph glands, one at each bronchial bifurcation, which act as a net to catch the migrating cells. These enlarged intrapulmonary glands can often be seen on

the cut surface of the lung at some little distance from the margin of the tumour.

The extrapulmonary glands involved are the parahilar, the inferior tracheo-bronchial and the lateral tracheal glands.

Lymph drainage

The lower lobes and the middle ones on the right side drain mainly into the inferior tracheo-bronchial, and the upper lobes into the lateral tracheal

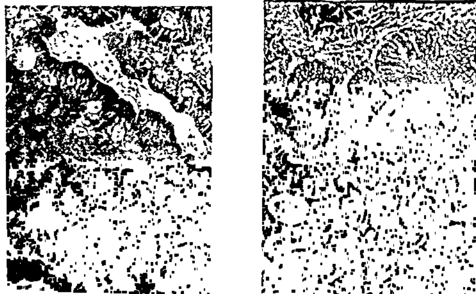


FIG 195.—Sections show columnar-celled adenocarcinoma.

group of glands. However, some lymphatics go to the alternative area and there is evidence to suggest that occasionally they may go to the area of the opposite side. A case is recorded of a contralateral paratracheal gland which was metastasized in the absence of any glandular involvement on the homo-lateral side. Secondary glandular involvement not infrequently occurs above the clavicle on the same side, and very rarely in the same position on the contralateral side. Axillary gland involvement is rare and most often occurs only when there is direct extension of the growth into the chest wall. Blood-borne metastasis occurs in the brain, liver, suprarenals, kidneys and bones. As would be expected the metastases most commonly reproduce the histology of the primary tumour, although they may differ.

2. CLINICAL ASPECTS

Clinical history

The clinical history is varied and can be broadly grouped under three headings

(1) History of acute respiratory infection

Those cases in which there is a history of an acute respiratory infection will be considered first. The onset is often acute, with a rigor and a clinical picture simulating that of pneumonia, and it is usually diagnosed as such. Treatment

by chemotherapy is usually effective. The patient, however, has a relapse in the ensuing weeks or months. This relapse should throw doubt immediately on the correctness of the diagnosis and further investigations should be carried out. In some cases, reaction to the primary treatment is not entirely satisfactory, in which case also the diagnosis should be questioned and further steps taken to elucidate it. If this possible mode of onset is borne in mind and any evidence that the course of the suspected pneumonia is atypical is appreciated, with the result that further investigations are instituted, many of the tumours in this group will be recognized earlier. Close questioning regarding the patient's condition prior to the acute onset will often elicit a history suggestive of the next group.

(2) Focal symptoms

Symptoms

Those presenting with focal symptoms are considered here. These are cough, haemoptysis, dyspnoea and chest pain.

(i) *Cough.*—This is most significant when it is persistent and non-productive. A cough of this type is most likely to be due to an endobronchial tumour.

(ii) *Sputum.*—Sputum may be present, however, and may be mucoid, mucopurulent or purulent in character. Even when the cough is accompanied by sputum but is persistent, it is worthy of investigation.

(iii) *Haemoptysis.*—About 60 per cent of patients have haemoptysis. This is very rarely severe, but is most commonly mere streaking, intimate staining or small clots.

(iv) *Dyspnoea.*—The majority of cases suffer from dyspnoea to a greater or lesser degree; this symptom is a significant one and coupled with any of the others should arouse a grave suspicion of a lung tumour.

(v) *Pain.*—This is a variable factor; it occurs in one form or another in about 60 per cent of cases. The pain is either retrosternal or located in the area of the chest wall over the growth, when it is due either to inflammatory or neoplastic involvement of the parietal pleura or the chest wall.

Retrosternal pain not infrequently occurs with growths which have not involved the pleura, either visceral or parietal, and has not been satisfactorily explained.

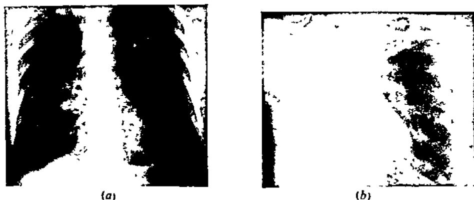
Persistent chest-wall pain should be investigated and the facile diagnosis of fibrositis or rheumatism should be withheld until it is firmly established that there is no underlying growth.

(3) General symptoms

Patients presenting with general symptoms with little or no focal manifestation are considered here. They often complain only of lack of energy, and this clinical group is on the increase.

Sometimes there may be a vague dyspeptic history, and in these cases, treatment has been directed to the alimentary tract, occasionally for long periods, often before the lung tumour has been diagnosed. Screening of the lung fields during barium-meal examination is easy and simple and will often produce valuable evidence in this type of case.

Some tumours are quite symptomless and are only recognized on mass radiography; more of these cases are coming to light since the institution of this method of examination. (Figs. 196 (a) and (b).)



(a) (b)

Fig. 196.—(a) Skiagram in course of mass radiography shows clear-cut circumscribed parahilar shadow. On bronchoscopy—tumour seen projecting into lower lobe bronchus. Biopsy showed squamous-celled carcinoma. Haemoptysis occurred for the first time after bronchoscopy. (b) Right pneumonectomy performed. Skiagram (after pneumonectomy) shows opacity of the right chest with fluid level at the extreme apex. Some cardiac and tracheal displacement to the right.

3. PHYSICAL SIGNS

Physical signs may cover the whole range of those found with pulmonary disease; they vary with the physical state of the underlying lesion.

In those cases associated with cough and sputum, râles and rhonchi are generally found, located in the affected lung or lobe.

Bronchial occlusion may give an asthmatic inspiratory or expiratory wheeze, depending upon whether the valvular action of the stenosis tends to be inspiratory or expiratory; the latter is more common. When the occlusion is complete, absent breath sounds and diminution of voice sounds are noted. Bronchial breathing and bronchophony are rarely met with, for their presence necessitates patency of the larger bronchi. When infection occurs behind the growth, fine and coarse râles, with perhaps signs of consolidation, may be present if the lumen is not completely occluded. In essence, carcinoma of the lung does not give rise to classical typical signs. There may be signs of fluid in the pleura which are quite classical. This in itself is evidence of pleural involvement by the growth and, in consequence, of inoperability. Aspiration and examination of the fluid with consequent finding of malignant cells will often establish the diagnosis.

4. SPECIAL INVESTIGATIONS

After the clinical history, the most important steps towards the establishment of a diagnosis are the special investigations which include radiography, bronchography, bronchoscopy, histological examination of a biopsy specimen if obtainable, punch biopsy and examination of the sputum for malignant cells.

(1) Radiography

In the first place, it cannot be too strongly stressed that a carcinoma of the lung may be present without any radiographic evidence of abnormality in the lung fields.

Carcinoma gives rise to variations from the normal by reason of the presence of a mass in the lung substance displacing air-containing tissue, or as a result of bronchial occlusion which alters the amount of air normally contained in the alveoli beyond the block. When there is a definite mass in the lung without bronchial occlusion, as is usual with a circumscribed tumour, the shadow is





FIG 197.—Five-months' history of bronchitis, loss of weight and lassitude. Small haemoptyses. Antero-posterior and lateral skiagrams show a large well-defined mass in right lower lobe lying in costo-vertebral sulcus. There is excavation with a fluid level. Note thickness of walls of cavity (had this been chronic lung abscess there would not have been the clear-cut definition of the mass, owing to surrounding pneumonitis). Right pneumonectomy performed. Tumour proved to be a squamous-celled carcinoma.

usually round or oval and reasonably well defined—less so than the remarkably sharp edge of the shadow of a hydatid cyst, but more definite than the rather hazy-edged shadow found with a lung abscess. These tumours, when degenerating, are sometimes diagnosed wrongly as lung abscess. The key to the problem is the relatively sharply defined edge of the shadow and the thickness of the wall of the cavity (Figs. 197 and 198); an abscess with walls as thick as these generally are would also have considerable surrounding haziness due to the associated pneumonitis. It is true that the rare case may have thin walls and still be a carcinoma; it is, however, extremely rare. If there is an associated bronchial block with a circumscribed growth, then the outline of the tumour may be completely lost in the surrounding atelectasis.



FIG. 198.—Same case as in Fig 197 Peripheral squamous-celled carcinoma showing marked degeneration and excavation of tumour. Note the clear-cut margins of the tumour, also the presence of several small deposits in other parts of the lobe.

(a) *Fluoroscopy*

Cases with incomplete bronchial occlusion may only be diagnosed by fluoroscopy, when it will be seen that the area supplied by the affected bronchus

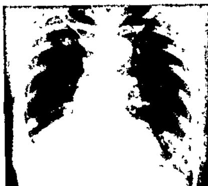




Fig 199.—Antero-posterior and lateral skiagrams showing collapse of lower and middle lobes Lateral film shows the migration of the upper lobe down into the costo-vertebral sulcus behind the lower lobe. History of right-sided pneumonia and pleurisy, 16 months previously Since then, cough with production of sputum which was occasionally blood-stained



Fig. 200—Same case as in Fig 199 Bronchogram demonstrates the block of the descending bronchus just below the upper lobe bronchus The level of the block has a convexity upwards which shows a slight irregularity in its contour. Bronchoscopy showed a soft reddish tumour obliterating the lumen of the descending bronchus below the level of the upper lobe bronchus Biopsy showed adenocarcinoma



Fig. 201.—Same case as in Fig 199 Skiagram shows post-operative condition—an opacity of the whole of the right side with tracheal displacement and slight displacement of the heart to the right.



Fig. 202.—Figure shows a section of the lung removed at operation There is a lobulated circumscribed tumour occupying the greater part of the lower lobe The projection of the tumour into the descending bronchus is also shown Note the small peripheral metastasis in the lower lobe

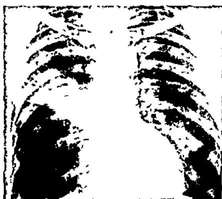




FIG. 203.—Five months previous to examination, the patient had double pneumonia. The right lung resolved quickly—the left lung more slowly. The patient returned to work but noticed steady loss of weight. Three weeks previous to examination, slight haemoptysis occurred. X-ray examination was then carried out. Antero-posterior film does not show any abnormality. Lateral film shows collapse of the anterior basic segment of the lower lobe. Bronchoscopy showed occlusion of the left anterior basic bronchus by a pink mass. Biopsy showed oat-celled carcinoma.



FIG. 204.—Same case as in Fig. 203. Skiagram after pneumonectomy shows marked displacement of trachea and migration of upper lobe to left side. Despite small size of growth, the patient died 14 months later with recurrence in mediastinum and cerebral metastasis.





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the antero-lateral segment of the right upper lobe. Bronchoscopy revealed a pink ... mass occluding the bronchus of the antero-lateral segment. Right pneumonectomy performed. Lung showed small whitish tumour 2 centimetres in diameter, completely occluding antero-lateral branch of the bronchus. Histology shows an adenocarcinoma.

does not light up as well as the rest of the lung field, or conversely that the affected area is much lighter. This may also be seen on the skiagram, indicating a localized area of emphysema. Most commonly this group gives rise to the classical picture of collapse of either a segment, or a lobe or occasionally of the whole lung (Figs. 199–205). When there is infection behind the growth, the affected area may appear to be bigger than normal, and occasionally a cavity with a fluid level may appear when there is excavation of the lung behind the growth, and rupture into a bronchus beyond the block has occurred. Skiagrams may also show the presence of a pleural effusion. At least one penetration x-ray examination should be made in order to show the bony framework, especially when pain is a prominent symptom, for not infrequently erosion of the ribs by the growth can be so recognized.

Fluoroscopy should always be carried out, if only to see whether the diaphragm is moving normally. Paralysis indicates mediastinal involvement and so renders the case inoperable.

(*b*) *Tomography*

Tomography generally speaking is not of great help. It will show widening of the inferior tracheo-bronchial angle, but this can be determined by penetrating films or by bronchoscopy.

(*c*) *Bronchography*

Bronchograms

In many cases the pathological cause of occlusion of the larger bronchus can be established directly by bronchoscopy; in those in which the site of the occlusion is beyond the range of bronchoscopic vision and in which it might be assumed that bronchography would be of value, this method does no more than establish the site of the block; the pathological cause is still undetermined.

Barium swallow

In any but the early cases, a barium swallow should be carried out. This will often show marked oesophageal distortion due to involvement of para-oesophageal glands, which does not necessarily indicate that the case is inoperable, since the glands sometimes can be removed, but still is an index of operative difficulty.

(2) Bronchoscopy

Bronchoscopy should be performed in addition to radiography as a routine investigation in all cases in which a carcinoma is suspected.

(*a*) *Anaesthesia*

The examination is carried out under local anaesthesia after pre-operative sedation with Omnopon $\frac{1}{3}$ grain and hyoscine hydrobromide $\frac{1}{150}$ grain. Five to ten minutes before commencing, the patient is given a gargle of amethocaine hydrochloride $\frac{1}{2}$ grain in 3 ounces of warm water.

Anaesthesia is produced by first anaesthetizing both vallecular fossae with 2 per cent amethocaine hydrochloride solution on gauze pledgets carried on special forceps (Krause's). An endotracheal injection of 10–15 minims of the same solution is given, the patient being postured to the side of the suspected growth after the injection. It may be necessary to reinforce the anaesthesia after the bronchoscope has been introduced, by spraying the solution into the bronchi with a long-nozzled spray. A table with a special head-rest is a

great help, but an ordinary table the head end of which can be lowered or elevated will serve quite well.

(*b*) *Technique*

It is not necessary to use a laryngoscope to introduce the bronchoscope, in fact in difficult cases it is impossible to use it. In consequence, it is better as a routine to use a bronchoscope without it and so master the technique on the easy cases.

The neck should be bent forwards with dorsiflexion of the head at the atlanto-occipital joint, as though "sniffing the air". Extension of the neck, by fixing the larynx against the spine, makes it immobile and renders elevation of the base of the tongue and epiglottis difficult or even impossible. The fingers of the left hand should hold the bronchoscope as it lies within the cavity of the mouth and should act as the fulcrum upon which to manipulate the instrument; this precludes that leverage on the teeth or gums about which the patient will complain most.

The bronchoscope is introduced until the tip of the epiglottis is seen; this is elevated by the instrument and the glottis is in view. The patient then phonates and movements of the cords are noticed. The bronchoscope is passed through the larynx and the subglottic region and trachea are inspected. Local bulging of the tracheal wall usually indicates enlargement of the paratracheal glands. Distortion of the trachea over a long curve usually towards the affected side is due to displacement only, occasioned by atelectasis. Distortions of the carina and tracheo-bronchial angles are of significance. The carina varies in normal individuals from literally a " knife edge", through "sharp", to a ridge about 1 millimetre broad; deformity, in its early stages, is really sub-carinal and is due to enlargement of the inferior tracheo-bronchial glands—later the carina itself may be broadened sometimes to such a severe degree as to encroach severely upon the lumina of both main bronchi. It can be taken that sub-carinal and, more so, carinal widening indicate that this group of glands is involved by growth; inflammatory glands do not distort this region but mould themselves round the bronchi. Unfortunately the converse is not true, lack of distortion of the sub-carinal region does not necessarily indicate that the glands are free from disease.

The bronchi are inspected, those of the lower lobes and of the middle on the right side by direct vision, and those of the upper lobes with a right-angled telescope.

The growth may be visible, appearing as a nodular mass, a nodular stenosis, or a soft vascular or greyish necrotic mass. Biopsy with microscopical examination of the specimen will clinch the diagnosis. A certain amount of experience is needed, however, for the correct interpretation of the histology of these fragments.

Often there is no growth in the lumen within telescopic vision; nevertheless distortion of one of the branch bronchi or one of the secondary carinas (in combination with the history and skiagrams) gives good deductive evidence as to the presence of a growth. When there is a distortional stenosis a blind biopsy will often give the desired evidence.

Aspiration of secretion examined as though it were sputum sometimes provides positive evidence.

Thus bronchoscopy may provide evidence of a tumour; of inoperability, due to either laryngeal paralysis, or carinal or sub-carinal widening or both, of the extent of main bronchus involved by growth, as an index of the feasibility of amputation of the bronchus clear of the growth; and lastly but equally important it may fail to show bronchoscopic evidence of inoperability.

(3) Examination of sputum

Examination of the sputum

In 1935, Dudgeon and Wrigley described a method of examining the sputum to demonstrate the presence of malignant cells—an elaboration of Dudgeon's wet-film method for the examination of tumours.

The sputum must be fresh, to avoid autolysis, and non-purulent; the more solid or blood-stained particles of the sputum are taken, and a thin film on the slide is fixed in Schaudinn's fluid, then stained with haemalum and counterstained with eosin. Considerable experience is needed to interpret the slides, but once the technique and experience are acquired it will be found invaluable in doubtful cases.

(4) Punch biopsy

Punch biopsy, in which a small cylinder of the growth is taken out with a specially devised punch, should be reserved for inoperable cases only, when confirmation of the diagnosis is desirable. In operable cases it is unjustifiable.

5. TREATMENT

There can be no question that at the present time resection of the lung with a growth gives to the patient the best chance of survival. X-ray therapy with modern technique and apparatus provides good palliation, and it may be that future development will so improve results as to warrant its being used as a first choice.

6. DECISION TO OPERATE

Choice of case

The following considerations determine the type of case to be submitted to operation.

(1) Contra-indications

Evidence of inoperability found during investigation has already been discussed. This evidence is summarized below.

(a) Paralysis of both the recurrent laryngeal and the phrenic nerves or of either alone.

(b) Bronchoscopic evidence of involvement of the inferior tracheo-bronchial and lateral tracheal glands.

(c) Extension of the growth in the main bronchus beyond the limit for satisfactory removal.

(d) Enlargement of the supraclavicular glands.

(e) Pleural involvement as indicated by an effusion.

(f) Signs of distant metastasis.

(g) Evidence of the Pancoast syndrome.

Local factors

The ultimate decision as to operability can only be made after thoracotomy. With experience, it has been found that an increasingly greater number of cases is suitable for operation. Thorough exploration of the growth, including the mediastinum itself, should be carried out before the case is abandoned.

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Involvement of the chest wall, diaphragm or pericardium unless it is extensive, does not contra-indicate resection since these structures can be removed with the growth. Involvement of the mediastinum, even though localized, does constitute evidence of inoperability, for experience with this type of case has shown that early recurrence is probable.

Large masses of parahilar glands, if removable, form no bar to operation, because in the majority of cases, the enlargement is inflammatory and not neoplastic in nature.

Age as such is not a contra-indication if the general condition, cardio-respiratory reserve and local conditions of removal are good. The length of the operation time is of considerable importance in the older age-group; if the operation can be completed in 2–2½ hours, then little change in the patient's condition is to be anticipated at the end of the operation. The oldest patient operated upon was 69, and the youngest 21 years of age.

Conditions which contra-indicate operation in other fields do so also in this field. Diabetes mellitus, when properly controlled, offers no bar to surgery and, rarely, striking improvement in the diabetes will result from resection of the tumour (Figs. 206 and 207).



FIG 206 —Two months previous to examination, patient experienced pain in the right chest and loss of weight, with some staining of the sputum with blood. Treated symptomatically until 1 week before admission. Polyuria was present at this time X-ray examination carried out. Antero-posterior film shows circumscribed shadow in the axillary part of the right upper lobe with some atelectasis of the apex Examination of the urine and blood established the diagnosis of diabetes mellitus.



FIG. 207.—Same case as in Fig 206 Lateral film shows shadow just above the hilum in the postero-lateral segment of the right upper lobe Bronchoscopy showed some distortion of the right upper lobe bronchus but no evidence of tumour. The patient at this time was pyrexial, with a temperature of 100 4° F. The diagnosis was carcinoma of the lung with diabetes Right pneumonectomy performed The tumour proved to be a circumscribed peripheral squamous-celled carcinoma. There was no evidence of infection of the lung tissue.

(2) Type of operation

The ideal operation is pneumonectomy for it is only in this way that the whole lymphatic area can be removed at the same time. In certain cases, however, lobectomy is to be preferred. This is so in cases in which the respiratory reserve is low, for example, with emphysematous patients, generally in the older age-group, in whom the tumour is of the circumscribed type and in whom on exploring the chest there is no evidence of glandular enlargement.

(3) Pre-operative care

It is essential for these patients to have a pre-operative period in hospital of about a fortnight. This gives the patient a complete rest, allows of time for preliminary blood transfusion if indicated, and for pre-operative breathing and postural exercises. During this period also a further bronchoscopy is carried out unless it has been done quite recently—for the bulk of the investigations may have been conducted in the out-patient department. An important aspect of the pre-operative stay is normally not stressed—getting used to the hospital routine and the establishment of confidence and a friendly relationship between patient and medical and nursing staff.

Breathing exercises

These are carried out pre-operatively and post-operatively in all cases. In doing them, the patient is taught to expand the chest against pressure, so that he uses both the bases and apices of his lungs; he is taught also to use his diaphragm fully, especially during the expiratory phase, so that the diaphragm ascends well within the chest. During this time he is also given arm movements to practise and is persuaded to correct any faulty posture while lying in bed.

The respiratory efficiency of the patient can be increased remarkably in this manner and, especially in the post-operative period, it gives him confidence in himself. At a very early stage—indeed in a matter of a few days—he ceases to look upon himself as an invalid.

It is advisable during this period to get the patient to use a B.L.B. mask at intervals in order that he may become accustomed to it before the post-operative necessity for its use arises.

Twenty-four hours before operation, penicillin, 30,000 units 3-hourly, is given intramuscularly, and sulphadiazine, 1 gramme 4-hourly, by mouth. These dosages are continued for 3–4 days post-operatively also.

7. OPERATION

(1) Anaesthesia

Anaesthesia is as important for the success of operative interventions as is technique. All types of anaesthesia have been used for intrathoracic operations, local, spinal, over-pressure endotracheal anaesthesia, and endotracheal anaesthesia with controlled respiration.

Physiological considerations

The danger to life of an open pneumothorax has probably delayed the inception of surgery in the thorax as compared with that in the abdomen.

The main factors leading to a fatal outcome are anoxaemia and carboxaemia and possibly reflex disturbances from the cardiac and pulmonary plexuses in the mediastinum.

Ill effects of open pneumothorax

The obvious effects of an open pneumothorax are:

(1) Collapse of the lung on the affected side.

(2) Paradoxical respiratory movements of the collapsed lung.

(3) To-and-fro movements of the mediastinum during each respiratory phase.

(4) Finally, the less obvious loss of the aspiration effect of the negative intrathoracic pressure on the large veins.

The first two embarrass efficient respiration.

The alveolar gas-exchange area is decreased as a result of the collapse [illegible]

the inspiratory phase.

Pulmonary effects

There is a pendulum swing of air from the collapsed lung into the expanded one during inspiration and from the sound to the collapsed side during expiration, this incidentally causing air to rush in and out of the pleural cavity, often producing a "sucking noise" (the common description of this type of open wound). These disabilities give rise to a degree of anoxaemia which, in a patient with a low vital capacity, may prove to be fatal.

Mediastinal effects

Mediastinal flutter resulting from this mechanism apparently produces its ill-effects from reflex disturbances set up in the cardiac and pulmonary plexus. It is possible to open the chest widely under local or spinal anaesthesia in a patient with a good vital capacity, with very little ill-effect other than a small preliminary fall of blood-pressure, so long as the respirations are quiet and steady. If, however, the patient starts coughing, the blood-pressure drops considerably and the general condition deteriorates; in other words this deterioration occurs when the mediastinum commences to move, for during quiet respiration with the patient lying on the sound side, the mediastinum remains practically immobile This observation suggests a nervous reflex although it is difficult to obtain direct proof of this.

Cardio-vascular effects

The lack of aspiration effect on the larger intrathoracic veins leads to inefficient diastolic auricular filling and, as a direct result of this, to a decrease in the ventricular output, at first compensated for by increase in the cardiac rate, but eventually accompanied by such a serious drop in blood-pressure as to lead to heart failure.

From the above considerations it will be obvious that local and spinal anaesthesia are practicable only for patients with a good cardio-respiratory reserve, in fact only in the relatively young age-group and, in consequence, in the majority of cases of carcinoma of the lung, anaesthesia of this type would be quite contra-indicated

General anaesthesia by the endotracheal route is the obvious choice. Crafoord (1938) first stressed the disadvantages of the Meltzer-Auer over-pressure method, and the importance and advantages of what he has termed rhythmic ventilation and what we call controlled respiration. The essential difference between the two techniques is that in the over-pressure method the gases are delivered into the trachea in a steady stream at a slight positive pressure, the respiratory movements being carried out by the patient's own respiratory mechanism. Crafoord showed experimentally in dogs that although the oxygen tension in the blood can be maintained at a satisfactory level with over-pressure anaesthesia, the volume per cent of CO_2 rose to 94·2 after three hours—the fatal level in the curarized animal; he also produced evidence of decrease in the alkali reserve, and of acidosis. He showed that in an animal with serious CO_2 accumulation from the over-pressure method rhythmic ventilation produces a dramatic fall in the concentration of CO_2.

The effect of rhythmic or controlled ventilation can be best appreciated when the chest is opened, in a case in which occlusion of the bronchi of the lung on the operated side has not been carried out. At each inspiratory phase the

mediastinum elevates and the lung on the operated side expands, and when the pressure on the bag is released the lung retracts and the mediastinum falls back. With the over-pressure method, when respiration is carried out by the patient's own mechanism, these movements are minimal and the mediastinum tends to remain in the inspiratory position, that is, displaced towards the opposite side.

There is no possible doubt that controlled ventilation is not only preferable to the old over-pressure method, but is absolutely essential for the safety of the patient in the older age-groups.

Modern thoracic anaesthetists depend upon cyclopropane as the main anaesthetic agent because a high percentage of oxygen (up to 90 per cent) can be used with it. A closed air-tight circuit is used, with a CO_2 absorption chamber incorporated. The combination of the anaesthetic agent and the high oxygen and low CO_2 tension in the blood makes automatic respiratory control a simple matter. It is as well to mention at this point that with this method of anaesthesia, especially in the older patients, cardiac irregularities, sometimes leading to ventricular fibrillation, may occur; they have been attributed to cyclopropane. Introduction of 10–15 cubic centimetres of 1 per cent Novocain into the pericardial sac when the irregularities commence, has proved to be very efficacious in their control.

Toxic effects

One of the operative risks is an aspiration of purulent secretions, if present, into bronchi of the unresected lung, with consequent pneumonia. Its prevention has been the subject of much thought. Magill (1934) first introduced endobronchial balloons in order to occlude the bronchus of either the lobe or the lung to be removed, and this still is the common practice, the most efficient being the Thompson balloon which is enclosed in a silk net which fixes the balloon firmly in the bronchus when it is blown up and prevents it from slipping.

Aspiration

Broncho-pneumonia

Pre-operative sedation with morphine $\frac{1}{4}$ grain or Omnopon $\frac{1}{3}$ grain and hyoscine hydrobromide $\frac{1}{150}$ grain is followed by anaesthetization of the pharynx, larynx and bronchial tree with 2 per cent amethocaine hydrochloride. The endobronchial balloon and endotracheal catheter are placed in position, unconsciousness is induced by an intravenous injection of Sodium Pentothal, and then full general anaesthesia is instituted. During the induction of the anaesthetic a drip blood transfusion is set up, either through a needle or through a cannula after exposing the vein. The latter method is preferable in any case in which it is anticipated that post-operative transfusion or infusion will be needed.

Pre-operative sedation

Blood transfusion

The necessity for having a transfusion going throughout the operation will be fully appreciated if it is realized that by mischance the pulmonary artery or one of the veins may be torn during the procedure. The blood loss in these cases is sometimes severe, but equally important is the fact that, as far as the left ventricle is concerned, it is lost on the intake side with a consequent serious drop in output and blood-pressure. This can be quickly rectified by speeding up the drip. If, however, transfusion is commenced only after such an accident, the delay of 15–20 minutes before fresh blood is introduced will usually lead to a delay of many hours in bringing the blood-pressure back to the level which obtained before the accident; if the blood loss at the time has been serious the delay may be fatal.

(2) Operative technique

The patient is placed on his side, the front of the chest being supported by a rest which should not rise much above the midline, since the incision must extend to the anterior ends of the ribs. The pelvis also should be supported anteriorly and posteriorly, otherwise the patient's position is liable to change suddenly, perhaps at a crucial moment. The upper arm is carried forwards and towards the head, so as to take the scapula upwards and forwards with it. The diathermy pad is strapped to the patient's leg.

Incision

After the skin has been sterilized and the towels are placed, the incision is planned, the site depending upon the rib to be resected. The ribs are counted and the anterior end of the incision through the skin is accurately placed over the selected rib.

Selected rib

The sixth rib is the most generally useful, but in upper lobe tumours the fifth is preferable, and when difficulty with the lower lobe is anticipated the seventh rib is sometimes chosen. Incision through skin and muscles over the sixth and seventh rib is direct, but the fifth rib is under cover of the scapula, even when the arm is well abducted, and consequently the skin and muscular incision must be carried from the level of the fifth rib anteriorly below the angle of the scapula up in the scapulo-vertebral angle towards the fourth dorsal spine. *This allows of adequate mobilization of the scapula, which can be carried out* of the way when the chest wound is spread by a modified De Quervain's rib-spreader.

The chest is entered through the bed of the resected rib. Care should be taken in elevating the periosteum to preserve it intact, for accurate closure of the chest depends upon this; sutures will cut out unless the periosteum is present to take the tension.

The whole wound is draped with towels and is then spread open manually, the retractor merely retaining the degree of spread. Spreading should be done [illegible] accident. [illegible] extends [illegible] over of the erector spinae.

The intrathoracic condition is assessed and if the case is operable pneumonectomy is performed.

Dissection of hilum

Sound anatomical knowledge of the hilar structures is essential and repeated post-mortem dissections are advisable before venturing upon a dissection of the hilum in the living patient. The order in which the structures are taken will depend upon the circumstances. Usually, the pulmonary artery is first secured. Its exposure is carried out after section of the pleura behind, above and in front of the lung root, preferably by sharp dissection with scissors The upper margin of the artery is defined, and then with the index finger (a much safer instrument than forceps) the posterior aspect can be freed. On the [illegible] dissected from the front of the artery, and [illegible] a triangular [illegible] ing branches, is dissected off the artery. This allows the front of the artery to be completely and easily exposed and allows of the proximal ligature being placed near the origin of the right branch. The lower margin of the descending artery is under

cover of the superior pulmonary vein, but with finger and thumb the artery can be pulled upwards to define its lower margin, and a forceps can be safely passed behind it. The artery is ligated proximally and distally, the distal ligatures being placed on both branches of the artery close to the lung with No. 20–25 linen thread. A good rosette of artery should be left beyond the proximal ligature to preclude slipping. On the left side, the proximal ligature is placed close to the ductus arteriosus.

After the artery has been divided, the bronchus is identified and freed and a bronchial clamp is placed in position. Both pulmonary veins are tied and divided in the same way as the artery. The next step is the dissection of the inner margin of the lung from the pericardium, which entails division of the ligamentum latum pulmonis in which recognizable vessels may have to be secured. The position of the bronchial clamp is now reviewed to see if it is well up to the level of the carina, and if so a second clamp is placed on the bronchus distally, and the bronchus is divided. The inferior tracheo-bronchial glands are removed with the lung. The clamp on the bronchus is now removed, to see whether or not it is close up to the carina, and is adjusted; if it is not, then any excess bronchus must be removed. Enough bronchus is left beyond the clamp to allow of the removal of the terminal cartilage, which is excised from the mucosa and fibro-muscular wall.

Bronchial closure

The main factors leading to satisfactory bronchial closure are:

(1) In lobectomy the bronchus of the resected lobe must be cut close to the bronchus of the remaining portion of the lung, and in pneumonectomy the bronchus cut close to the carina, so that no pocket is left in which infected secretions can remain to seep along the suture punctures and start a peribronchial infection, which in turn causes either an empyema, or a bronchial fistula, or both.

(2) Suture of the end of the bronchus must be air-tight and must remain so for 10–14 days.

(3) The peribronchial tissue covering the stump is the ultimate factor upon which sound bronchial healing depends The tissue of the bronchus itself does not heal very well and a sound cover is the best insurance of success.

If these three principles are followed, the variations in technique and in the type of suture material are of little importance

The practice of removal of the cartilage ensures soft supple tissue for suture and the placing of longitudinal mattress sutures round the clamp, which are firmly but not tightly tied after the clamp is removed, tends to take tension off the terminal suture and so satisfies the second prerequisite. A pleural flap is sutured over the stump on the right side; on the left side, the stump is so buried in the mediastinum under the aortic arch that cover is automatically provided when the patient is turned over on his back and the heart rotates to the left, thus closing the mediastinal gap.

The pulmonary artery stump is now tucked into the pocket of pericardium, made during the freeing of the artery, with a few catgut sutures. This keeps the arterial stump away from the bronchial stump and eliminates the risk of a secondary haemorrhage, should there be any peribronchial suppuration.

Penicillin 50,000–100,000 units either as a solution or with sulphathiazole powder is put into the pleura and the chest is closed without drainage.

Before the last intercostal suture is tied a pair of forceps holds the space open and the anaesthetist inflates the lung to its fullest extent; this elevates the mediastinum which otherwise would be depressed and thus ensures that, at the end of the operation, the mediastinum is centrally placed.

The hemithorax is then firmly strapped. Without this the patient finds it difficult to cough owing to lack of support. Preferably the patient is transferred to his own bed, while in the theatre, and is kept recumbent until he is conscious. Then he is gradually raised to the sitting position. Of course, this move is delayed if there is any cardiovascular insufficiency. Continuous oxygen is given, and should be administered freely in the immediate post-operative period especially when any nursing procedures are necessary.

8. POST-OPERATIVE CARE

The patient is nursed in a sitting position for the first 3 weeks. This position allows of more efficient diaphragmatic movements and, moreover, fluid which accumulates in the pleura remains at the bottom of the sac.

Post-operative complications

Certain complications may have to be treated. The chief of these are aspiration pneumonia, bronchial fistula and infection of the pleural space.

Pneumonia results from aspiration of secretions during the operation as already indicated, and also from retention of secretions in the bronchi post-operatively. In consequence, it is essential to encourage the patient to cough up his secretions—a painful procedure. Morphine ¼ grain should be given every 6 hours to control the pain if necessary for the first day or so, and the chest on the operated side should be firmly supported by the attendant nurse. It is also an encouragement to the patient to assure him that though it is a painful procedure he can do himself no harm in coughing, and that it is essential for him to cough up secretions when they are present.

With a portable apparatus, skiagrams are taken of the chest 24 hours or less after the operation, especially so if there is any respiratory distress. These will not only demonstrate the state of the remaining lung but also the position of the mediastinum which should be central; if it is not so, air should be aspirated or introduced according to requirements.

The pleural cavity eventually fills with fluid, but it is well to keep the fluid below the level of the hilum until after the first fortnight when presumably the bronchus will be soundly healed. If this precaution is not taken and the bronchus does leak, the patient is liable to be drowned by the aspirated fluid.

If there is any rise in temperature, the fluid should be aspirated and examined bacteriologically. There may be loculation of the fluid, indicated radiologically by multiple fluid levels. It will be necessary when there is any pyrexia to aspirate each loculus for examination. If the loculus is above the hilar level its content must be removed by aspiration. Penicillin, 30,000–50,000 units, is injected after each aspiration even in the apyrexial cases.

When frank infection has occurred, aspiration and penicillin instillation can be tried if the organisms are penicillin-sensitive. When there is a fistula, it is safer to institute dependent drainage, either with an intercostal tube or after rib resection.

Fistulae will most often close spontaneously at a later date, and then vigorous sterilization of the pleura can be undertaken; when it is sterile the tube

can be removed. The common organisms other than the penicillin-sensitive group are the coliform group—these can be eliminated by introducing a 10 per cent solution of a proprietary preparation U.F.I.

Patients are kept in bed for from 4 to 6 weeks; the older the patient the longer the post-operative rest in bed. This seems to be warranted to ensure a period for cardio-respiratory adjustment. In the older age-group, auricular fibrillation may occur, generally however, not until the second or third day It can be controlled with digoxin or digitalis.

9. RESULTS

Of the total number of cases seen it can be anticipated that only about 15 per cent will be deemed suitable for operation. Of those operated upon, in 60 per cent only will pneumonectomy or lobectomy be possible.

The results in 153 of my own cases submitted to operation were as follows:

Pneumonectomy	89	Operative deaths	20
Lobectomy	4		
Thoracotomy	60	Operative deaths	8

These results include all the cases dealt with since pneumonectomy was first utilized as a method of treating carcinoma of the lung when the technique had not been standardized Since that time indications of operability have been considerably extended, and today, cases which hitherto were considered hopeless are being submitted to extensive operative procedures.

In 5 cases large portions of the pericardium have been resected. In 2 cases part of the auricular wall, in 2 cases portions of the chest wall, and in 2 cases portions of the diaphragm were similarly excised.

The operative mortality includes all those patients who were not able to leave hospital after operation and is just over 20 per cent for both thoracotomy and pneumonectomy.

Eighteen cases have survived more than 1 year, 9 more than 2 years, 4 more than 4 years, 3 more than 5 years, and 1 patient is still alive and well 10½ years after operation. Twenty-six cases are alive less than 1 year after and 7 cases have not yet been traced.

It can be anticipated that the mortality figures eventually will be less than 10 per cent and time alone will determine whether or not the extensive resections practised today will be justified by the late results.

PART II
ADENOMA OF THE BRONCHUS

GENERAL

Adenoma of the bronchus is the second most common tumour of the lung, forming 6 per cent of the total, occurring more commonly in women than in men in the proportion of 4 : 1.

(1) Site of origin

This tumour most frequently arises in the larger bronchi, that is, the main, lobar or segmental bronchi; occasionally, however, tumours occur in the more peripheral portions of the lung, arising from the smaller bronchi, and more rarely in the trachea itself. It is generally accepted that the tumours have their origin in the bronchial glands. These are sero-mucous glands containing crescents of Gianuzzi, in fact they are sero-mucous salivary glands in miniature with ducts opening on the surface of the bronchial mucosa. The glands themselves lie in the submucosa, and often a portion of the gland may project between the cartilaginous rings, lying outside the lumen of the bronchus. The tumour may be purely endobronchial, arising from the bronchial wall by a broad base or by a relatively small pedicle. It may be both intrabronchial and extrabronchial, however, and often the extrabronchial mass may be many times larger than the intrabronchial portion. This distribution can be understood easily from the normal anatomical arrangement of the gland. In the more peripheral type as far as it is possible to ascertain, the tumours are extrabronchial only.

Bronchial glands

Macroscopically, the tumours are sharply circumscribed, firm in consistency and whitish in colour.

Macroscopical appearance

(2) Histology

Histologically, the striking feature is the regularity of the cells, which are cuboidal with well-staining nuclei. The cells are arranged either in acinar formation, in columns or in masses; one or other arrangement normally dominates the picture, although all three arrangements may be present in the same section. Occasionally there is an excessive secretion of mucin, giving a picture reminiscent of the mixed salivary tumours. It is impossible to distinguish the usual histological picture from that of the non-mixed type of salivary tumour. The connective tissue of the tumour is of the adult type and contains numerous blood-vessels. There is also a well-defined capsule and, when the tumour is intrabronchial, it is covered by the bronchial mucosa, which may be of the normal type or of the squamous metaplastic type. The tumours are generally benign, but occasionally they may take on locally invasive features of the basal-celled carcinomatous type. They have also been described as undergoing true carcinomatous change, giving rise to glandular metastasis. In one case of Roberts's, a glandular metastasis was found, and both this and the primary tumour were histologically benign.

Symptoms

The tumour produces its symptoms either by ulceration, giving rise to haemoptysis, or by causing bronchial obstruction with or without infection

(3) Clinical signs

The symptoms and clinical signs are similar to those occurring with carcinoma of the lung. Usually, however, there is not the same degree of toxaemia, and the history is generally long, often being of years' duration.

(4) Diagnosis

(a) *Bronchoscopy*

Diagnosis is established by bronchoscopy for, in the majority of cases, the tumour lies within telescopic vision and, except for a proportion arising in the segmental branches of the upper lobe bronchi, biopsy of the tumour is possible, and a definite histological diagnosis can be made.

The tumour presents as a smooth cherry-red rounded swelling covered by normal mucosa, unless ulceration has occurred when the surface is necrotic. The tumour does not bleed easily when touched, but not infrequently after biopsy there is free bleeding which is explained by the vascularity of the growth. The bleeding stops quickly and is never sufficient to cause anxiety.

In the rarer peripheral groups, the diagnosis is made upon deductive grounds from the radiological evidence.

(b) *Radiography*

Plain skiagrams may show atelectasis of the lung, lobe or segment, but as with carcinoma of the main bronchus type, there may be no radiographic evidence when bronchial occlusion has not occurred.

Bronchography may be of assistance in the latter group, showing a clear-cut filling defect in the bronchial outline. The crescentic outline of the bronchial block and its possible occurrence with carcinoma has already been noted, but this appearance is more common with adenoma.

(5) Treatment

Current treatment can be considered under the following headings:

(a) Local removal of the tumour.
(b) Irradiation therapy.
(c) Lung resection.

(a) *Local removal*

Local removal of the tumour through a bronchoscope is practised in many American clinics, and in carefully selected cases gives satisfactory results. This method, however, is only possible when the growth is purely intra-bronchial, and even when this is the case, recurrence is seen in a percentage of cases. This method also does not take cognizance of changes occurring behind the tumour.

(b) *Irradiation*

Irradiation therapy has been used both by local implantations of radon seeds and by deep x-rays. These tumours are radio-sensitive, as would be expected when their similarity to the non-mixed type of salivary tumour is considered. This method of treatment, although dealing with the intrabronchial and extrabronchial portions of the tumour, does not deal with any pulmonary change that occurs beyond the growth.



FIG. 208 —The patient had pneumonia at the ages of 10, 11, 13 and 16 years. One month previously had haemoptysis and occasional cough productive of ½ oz. of mucoid sputum Some dyspnoea on exertion. Bronchoscopy showed smooth rounded tumour opposite the right upper lobe bronchus. Biopsy—histology—adenoma of the bronchus Right pneumonectomy performed. Lower and middle lobes densely adherent to chest wall, mediastinum, diaphragm and upper lobes Larger masses of parahilar glands. Hilum dissected with great difficulty owing to glands and intrahilar fibrosis. Skiagram shows collapse of lower and middle lobes





FIG 209.—Same case as in Fig 208 Right bronchograms show filling of the upper lobe bronchus, with complete block of the descending bronchus at level of upper lobe, exhibiting a smooth convexity upwards



FIG. 210 —Same case as in Fig 208. Post-operative skiagram shows uniform



FIG. 211.—Same case as in Fig 208 Lung removed showing intra-bronchial and extrabronchial portions of the tumour with gross bronchiectasis and shrinkage of the lower and middle lobes, the upper lobe being relatively normal.

(c) Lung resection

The present tendency, in Great Britain at least, is to subject all cases of adenoma of the bronchus to lung resection by either lobectomy or pneumonectomy. It is felt that the two factors referred to above, the occurrence of an extrabronchial mass, and the changes in bronchi behind the tumour in such a

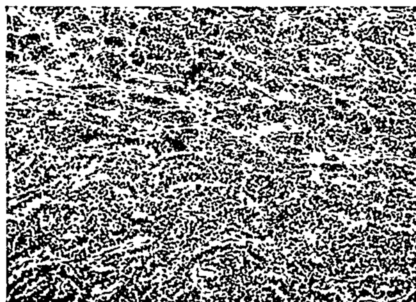


FIG 212 —Histology of the tumour illustrated in Fig 211. The attempt at acinar formation in this case is primitive, the cells being arranged rather in columns and clumps.

high percentage of cases, make the less radical measures unsatisfactory. (Figs. 208–212.) Added to this is the good prognosis both as to mortality and morbidity, which makes lung resection the best method at the moment. It is possible that with greater experience the type of case which can be treated satisfactorily by more conservative means will be recognized more easily.

PART III
OTHER TUMOURS

Other tumours of the lung are rare. They are hamartomas and angiomas.

1. HAMARTOMAS

Hamartomas are tumours which are predominantly composed of cartilage, but they also contain epithelial elements, fat and often muscle fibre, in other

words, all the elements found in a normal bronchus but having no regular arrangement. It is probably correct that true chondromas of the lung do occur, but many of these have subsequently proved to be hamartomas. The tumours occur in the substance of the lung as circumscribed lobulated tumours, showing on section a firm whitish appearance, predominantly cartilaginous, often with cystic spaces and sometimes with well-marked calcification and sometimes ossification.

Occasionally chondromatous intrabronchial tumours are found. These are probably true chondromas, but may possibly fall into the hamartoma group despite the present lack of evidence of mixed histology. There is no evidence to date that these tumours become malignant.

Symptomatology

Clinically they give rise to very few symptoms and are most often found either by routine x-ray examination of the chest or during radiological examination of the chest for some other respiratory affection.

The intrabronchial tumours give rise to symptoms of bronchial obstruction. When the tumour is pedunculated the characteristic cough is produced, which is spasmodic in type almost like whooping-cough; if there is no infection of the bronchi, there will be no sputum. Generally, however, the cough is productive. Bronchoscopy in this group establishes the diagnosis.

Radiology

The radiographic appearances are those of a sharply demarcated shadow, oval and lobulated in shape and sometimes showing stippled calcification.

Treatment

Peripheral group

The tendency in the peripheral group is to perform pulmonary resection despite the lack of clinical symptoms or the risk of malignant change. Many cases are not diagnosed until after resection but, even in those cases in which a diagnosis is made, the uncertainty and the possibility of the diagnosis being wrong outweighs other considerations.

Intrabronchial group

The intrabronchial tumours are treated by bronchoscopic removal—if the whole tumour is not removed it will recur and necessitate further intervention.

2. ANGIOMAS

Angiomas of the lung occur in two groups, either as a cavernous angioma or as an arterial angioma.

(1) Cavernous angioma

These tumours are like cavernous angiomas in other parts of the body.

They may or may not be associated with angiomas in other parts of the body, such as capillary angioma of the skin, or of the mu[illegible] membranes of the mouth, pharynx, trachea or main [illegible]

Clinically they present with haemoptysis, o[illegible]
the patient remaining perfectly [illegible]

Radiographically, there may be [illegible]
as a circumscribed or conv[illegible]
evidence of capillary naevi in [illegible]
tract may afford a clue to the [illegible]
may not be arrived at before [illegible]

Treatment

Generally speaking, treatment is conservative, first because it is difficult to establish the origin of the bleeding, secondly because there is a distinct possibility that the lesions may be scattered throughout the whole of one or both lungs. If the origin of the bleeding can be established, and there is a radiographic shadow in the lung field even though there is no evidence of superficial angiomas, obviously pulmonary resection is the correct treatment.

(2) Arterial angioma

This group, which can be described perhaps better as a congenital arteriovenous aneurysm, is being more frequently recognized.

The cases that have been described usually have an aneurysmal dilatation, into which open both artery and vein—generally those supplying and draining the segments of the lobes.

Clinically they may present with haemoptysis or they may be recognized by their radiographic appearance, or by the typical "machinery" murmur most often heard over them; this murmur may only be present during the inspiratory phase, becoming a soft systolic souffle at the end of expiration.

Radiography shows a shadow of a rounded shape, and generally the large vessels entering and leaving the shadow can be seen; fluoroscopy reveals pulsation in the tumour, and if chymographic skiagrams are taken the pulsations can be verified.

Treatment

Treatment entails pulmonary resection, either segmental or lobar, for theoretically there is the risk of rupture into the pleural cavity. The wall of the aneurysmal sac is sometimes so thin that blood can be seen swirling in the sac when the chest is open, giving the impression that a sharp blow upon the chest wall might easily rupture it.

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[References to other titles are given under Lung—Tumours, in the Index Volume. The subject of Lung Diseases: Tumours, is also dealt with under the heading of Lung Diseases in the *British Encyclopaedia of Medical Practice* (1938), Vol. 8, p. 224.]

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Clinically they present with haemoptysis, often repeated over long periods, the patient remaining perfectly well in the intervening periods.

Radiographically, there may be little evidence of the tumour or it may appear as a circumscribed or convoluted shadow in the lung field. Bronchoscopic evidence of capillary naevi in the mucous membranes of the respiratory tract may afford a clue to the diagnosis. Failing such evidence, a diagnosis may not be arrived at before the lobe is resected.

Treatment

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[References to other titles are given under Lung—Tumours, in the Index Volume. The subject of Lung Diseases: Tumours, is also dealt with under the heading of Lung Diseases in the *British Encyclopaedia of Medical Practice* (1938), Vol. 8, p 224]

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Occasionally chondromatous intrabronchial tumours are found. These are probably true chondromas, but may possibly fall into the hamartoma group despite the present lack of evidence of mixed histology. There is no evidence to date that these tumours become malignant.

...mptoma-...logy Clinically they give rise to very few symptoms and are most often found either by routine x-ray examination of the chest or during radiological examination of the chest for some other respiratory affection.

The intrabronchial tumours give rise to symptoms of bronchial obstruction. When the tumour is pedunculated the characteristic cough is produced, which is spasmodic in type almost like whooping-cough; if there is no infection of the bronchi, there will be no sputum. Generally, however, the cough is productive. Bronchoscopy in this group establishes the diagnosis.

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Treatment

...ripheral ...oup The tendency in the peripheral group is to perform pulmonary resection despite the lack of clinical symptoms or the risk of malignant change. Many cases are not diagnosed until after resection but, even in those cases in which a diagnosis is made, the uncertainty and the possibility of the diagnosis being wrong outweighs other considerations.

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2. ANGIOMAS

Angiomas of the lung occur in two groups, either as a cavernous angioma or as an arterial angioma.

(1) Cavernous angioma

These tumours are like cavernous angiomas in other parts of the body. They may or may not be associated with angiomas in other parts of the body, such as capillary angioma of the skin, or of the mucous membranes of the mouth, pharynx, trachea or main bronchi.

Clinically they present with haemoptysis, often repeated over long periods, the patient remaining perfectly well in the intervening periods.

Radiographically, there may be little evidence of the tumour or it may appear as a circumscribed or convoluted shadow in the lung field. Bronchoscopic evidence of capillary naevi in the mucous membranes of the respiratory tract may afford a clue to the diagnosis. Failing such evidence, a diagnosis may not be arrived at before the lobe is resected.

Treatment

Generally speaking, treatment is conservative, first because it is difficult to establish the origin of the bleeding, secondly because there is a distinct possibility that the lesions may be scattered throughout the whole of one or both lungs. If the origin of the bleeding can be established, and there is a radiographic shadow in the lung field even though there is no evidence of superficial angiomas, obviously pulmonary resection is the correct treatment.

(2) Arterial angioma

This group, which can be described perhaps better as a congenital arteriovenous aneurysm, is being more frequently recognized.

The cases that have been described usually have an aneurysmal dilatation, into which open both artery and vein—generally those supplying and draining the segments of the lobes.

Clinically they may present with haemoptysis or they may be recognized by their radiographic appearance, or by the typical "machinery" murmur most often heard over them; this murmur may only be present during the inspiratory phase, becoming a soft systolic souffle at the end of expiration.

Radiography shows a shadow of a rounded shape, and generally the large vessels entering and leaving the shadow can be seen; fluoroscopy reveals pulsation in the tumour, and if chymographic skiagrams are taken the pulsations can be verified.

Treatment

Treatment entails pulmonary resection, either segmental or lobar, for theoretically there is the risk of rupture into the pleural cavity. The wall of the aneurysmal sac is sometimes so thin that blood can be seen swirling in the sac when the chest is open, giving the impression that a sharp blow upon the chest wall might easily rupture it.

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Radiology

The radiographic appearances are those of a sharply demarcated shadow, oval and lobulated in shape and sometimes showing stippled calcification.

Treatment

Peripheral group

The tendency in the peripheral group is to perform pulmonary resection despite the lack of clinical symptoms or the risk of malignant change. Many cases are not diagnosed until after resection but, even in those cases in which a diagnosis is made, the uncertainty and the possibility of the diagnosis being wrong outweighs other considerations.

Intrabronchial group

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2. AETIOLOGY

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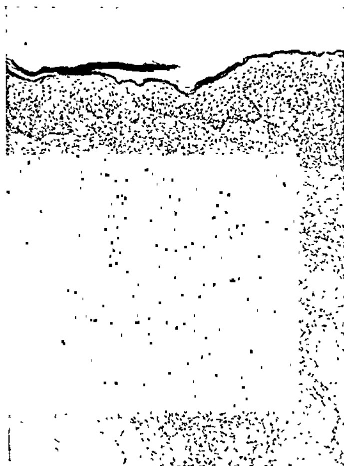


FIG. 213—Biopsy, 15th February 1947, before starting treatment. A tuberculous focus (× 90) (*Brit. med. J.*)

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LUPUS VULGARIS

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1. DEFINITION

226.] Lupus vulgaris is the name given to a variety of tuberculous infection of the skin of which the characteristic lesion is a small nodule known as a lupoma. This lesion may vary in size from that of a pinhead to a lentil. It is nearly flat or moderately raised above the surface, and reddish-brown in colour. Under glass pressure it appears as a brownish-yellow opalescent spot resembling apple jelly. The disease is gradually progressive, peripheral extension taking place by the formation of fresh nodules.

2. AETIOLOGY

Lupus occurs more commonly in temperate and cold countries than in warm sunny climates. In Australia, for example, it is very rarely seen. It is met with at all ages, but it begins in childhood and in adolescence far more often than in adult life. Poverty and defective nutrition and hygiene are common aetiological factors. The skin may be infected by external contact, but more frequently by the lymphatic route from a mucous membrane, especially of the nose, and from contiguous or neighbouring tuberculous glands, and exceptionally from underlying tenosynovitis or from disease of bone. When established in the skin the infection spreads by the lymphatics, fresh nodules appearing usually in the immediate neighbourhood of the early sites but

Climate

Age

Mode of infection and spread



FIG. 213.—Biopsy, 15th February 1947, before starting treatment. A tuberculous focus (× 90) (*Brit. med. J.*)

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In the type known as disseminate lupus the infection is blood borne. Lupus is usually unassociated with demonstrable tuberculous change in remote organs, but a *quiescent pulmonary focus* of infection may be present and the patient may die ultimately of pulmonary tuberculosis. In the disseminate type

an internal focus must be present though often it cannot be detected. When *there is demonstrable* tuberculosis of tissues other than those in the immediate neighbourhood of the cutaneous infection, it is usually found in bone or joints.

Occurrence in children on buttocks and lower limbs

In young children in whom the skin may be infected by contact with the floor, lupus is found with some frequency on the buttocks or on a lower limb. In the latter situation advice is often sought on account of suppurating tuberculous glands in the inguinal region, the cutaneous lesion, which may be quite small, having been overlooked.

3. MORBID ANATOMY

The lupus nodule feels firm to the touch but it is in reality a granuloma of rather soft consistency, which can be easily penetrated by a pointed match

Microscopical section

Microscopically the changes found are similar to those of tuberculosis of other organs (Fig. 213). The basic reaction is a proliferation of endothelial cells which in places undergo degenerative change but seldom reach that of complete caseation; giant cells may be present but generally in small numbers. To this characteristically tuberculous reaction may be added an inflammatory infiltration composed chiefly of lymphocytes. There is no characteristic epidermal change, though thinning or absence through ulceration are common secondary changes.

4. CLINICAL PICTURE

Common sites

Lupus is far more often seen on the face, neck and ears than elsewhere, but it occours also on the trunk, particularly on the upper part, the buttocks and the limbs. It is rarely seen on the genitalia or on the scalp. The tendency is towards a gradual centrifugal spread of the process by the formation of fresh nodules, accompanied sometimes by ulceration, while the older part of the lesion undergoes partial healing by cicatrization. Resolution, however, is rarely complete and active nodules are generally found in the scar tissue, a point which serves to distinguish lupus from superficial tertiary syphilis in the scars of which active disease is practically always absent.

There are several clinical types of lupus for which the following descriptive terms may be employed.

(1) Agminate lupus

Clinical types

This is the common type in which the lesion consists of a number of more or less closely aggregated lupomas, spreading excentrically and accompanied by central, rather dense scarring.

(2) Ulcerative and mutilating lupus

Ulceration and destruction of underlying tissue such as the cartilage of the nose, and mutilation of other affected parts are the special features of this variety. The progress of this destructive type may be moderately rapid.

(3) Scrofulodermia



(a)



(b)



(c)



(d)



(e)



(f)

(a)—Elephantiasic lupus of the left arm, before calciferol had been used The paler patches on the back of the shoulder and the trunk show results of local treatment, particularly acid nitrate of mercury

(b)—Lupus of the elbow after treatment with the Kromayer lamp and brass paste, showing how the lupus nodules are picked out.

(c)—Fungating lupus of the nose and ear.

(d)—Same patient as in (c), after local treatment for nearly 2 years. Calciferol was not then available.

(e).—Lupus of the mouth after treatment, showing microstomia.

(f)—Same patient as in (e), after plastic surgery.

PLATE V

(4) Elephantiasic lupus

The lymphatics of the lips, particularly of the upper lip, or of a limb are invaded by the process, with gross resultant elephantiasis-like swelling.

(5) Erythematoid lupus

This is seen most often on the cheeks and nose. The lupus is very superficial, accompanied by erythema and sometimes by scaling. There may be a close resemblance to lupus erythematosus.

(6) Verrucous tuberculosis (verruca necrogenica)

This variety, most often seen on the hand or wrist, and rarely about the anal orifice, is caused by direct inoculation of the infection into the skin sometimes from an external source, as in butchers or in post-mortem room attendants (verruca necrogenica, anatomical wart). In such, the primary lesion is a pustule which will not heal and which becomes transformed gradually into a granuloma surmounted by wart-like excrescences; it tends to spread centrifugally. This lesion may be followed by lymphangitis and tuberculous adenitis of regional glands. Warty tuberculosis, however, is perhaps more often seen in patients suffering from chronic pulmonary tuberculosis, in whom the infection has been implanted on the hand from the mouth.

Direct inoculation

(7) Lupus tumidus

The term lupus tumidus is used for cases in which the granuloma is raised well above the surface.

(8) Vegetating or fungating lupus

In this, exuberant vegetations are the striking characteristic of the infection or of a part of it.

(9) Lupus of mucous membranes

The gums, lips, palate, buccal mucosa, pharynx and nose may be attacked. On the gums the lesions consist of exuberant bud-like vegetations; elsewhere aggregation of vegetations may produce a mamillated or, by cicatrization, a reticular appearance; the lesion may ulcerate in parts. Perforation of the nasal septum is common. Blockage of a lacrimal duct is revealed by epiphora.

(10) Lupus of the conjunctiva

In this situation lupus is fortunately rare. A red granular change is the characteristic appearance, with a tendency to adhesions. The cornea is generally not affected.

5. EVOLUTION AND COMPLICATIONS

Untreated lupus persists indefinitely. Its rate of extension varies but is generally very slow. It is never directly the cause of death, but pulmonary tuberculosis may occur as a terminal event. Ectropion is a relatively common result of extensive lupus of the face; it may be followed by keratitis and dense corneal opacity.

Of the complications of lupus, epithelioma, either basal-celled or more often squamous-celled, is the most important. It has occurred, but by no means invariably, in patients who, in the past, have been treated by x-ray irradiation.

Cutaneous horns occur, sometimes in numbers, on the face and neck, in the scars of a few patients.

6. DIFFERENTIAL DIAGNOSIS

(1) Sarcoid

Sarcoid of Boeck may closely resemble lupus, especially the disseminate type. The diagnosis may be settled by biopsy and by the discovery of clinical evidence of sarcoid in lymph glands or in bones, or of the characteristic radiographical change in the lungs.

(2) Lupus erythematosus

The differentiation of erythematoid lupus and lupus erythematosus may also be difficult, and the diagnosis may have to be settled by biopsy. The histological changes characteristic of tuberculosis are never found in lupus erythematosus.

(3) Leishmaniasis cutis

Cutaneous leishmaniasis of the face and lupus may be absolutely similar, and here the diagnosis must depend upon the demonstration of Leishman-Donovan bodies in the lesion, either by culture or by section.

(4) Tertiary syphilis

Tertiary syphilis of the skin may also be quite indistinguishable from lupus both clinically and histologically but with very few exceptions the process is far more rapidly extensive in syphilis than in lupus. Moreover, active nodules are almost always found in the scar of lupus, whereas evidence of activity in that of tertiary syphilis is extremely rare. Nevertheless, sero-diagnosis is often required and, in the event of a negative reaction, a therapeutic test with potassium iodide should be made.

7. GENERAL TREATMENT

(1) Dietetic and physical methods

Nutrition
Actinotherapy
Diet

Until recently the general treatment of lupus consisted chiefly of the maintenance of a high level of nutrition supplemented by cod-liver oil and combined, when possible, with heliotherapy or with general carbon-arc-lamp baths. An advance in nutritional therapy was achieved by a certain type of dietary known as the Sauerbruch-Herrmannsdorfer-Gerson diet, which consisted essentially of a high proportion of uncooked food rich in vitamins, particularly of fresh meat, milk, cream and butter, raw vegetables and supplementary cod-liver oil. Heliotherapy was instituted on a large scale by Rollier in Switzerland; the same method was adopted in England chiefly in the treatment of children with lupus and surgical tuberculosis of other types, by Gauvain at Alton. In winter the carbon-arc lamp has been used to replace heliotherapy; this has appeared to be less effective than exposure to natural sunlight in which the stimulus to metabolism of exposure to fresh air is added to the specific effect of light. In all these measures the common factor is an abundance of vitamin D.



(*a*)



(*b*)



(*c*)



(*d*)



(*e*)



(*f*)

(*a*).—Patches of agminate lupus with actively growing edges and commencing fibrosis in the centre.

(*b*).—Ulcerative lupus, affecting the whole face Ectropion had already led to some corneal ulceration on the right side. A new upper eyelid was made to preserve as much of the sight as possible.

(*c*).—Vegetative lupus of face

(*d*).—Same patient as in (*c*), after calciferol and local treatment for 2 months

(*e*) —Lupus of the hands, associated with sinuses from multiple tuberculous dactylitis

(*f*).—Same patient as in (*e*), after calciferol and local treatment for 4 months

PLATE VI

(2) Treatment with calciferol

That vitamin D, combined with an adequate nutritional hygiene, is in fact all that is required has been amply demonstrated by the striking effect of the administration of irradiated ergosterol—crystalline vitamin D_2 (calciferol) in large doses. The drug is more rapidly effective when given by mouth than by intramuscular injection, though either route may be used. By mouth, the solution in propylene glycol, or tablets (Ostelin High Potency tablets), or capsules containing calciferol in oil, or an emulsion of the oily solution may be given. For injection, a solution in oil containing 15 milligrams or 600,000 international units per cubic centimetre may be used. In France, the standard treatment for otherwise healthy subjects (Charpy, 1948) consists of the administration by mouth of calciferol in alcohol and the dosage employed is 15 milligrams, 2 doses being given each week for the first 4 weeks, and 1 dose every week or 10 days during the ensuing months, up to a year or more if necessary. In England (Dowling and Thomas, 1946) the drug most often used has been the Ostelin High Potency tablet, each of which contains 1·25 milligrams of calciferol or 50,000 i.u., and the dosage employed has been 3 of these tablets daily for the first 2 or 3 months reduced to 2 daily when manifest improvement has set in. To children of 6 years or over, 100,000 i.u. daily may be given in the initial weeks.

Methods of administration

The effect of calciferol on lupus is to produce, sometimes after an early increase in the inflammatory aspect of the disease, a progressive flattening of the lupomas and, in some cases, their ultimate disappearance.

Effect of calciferol

Close observation of a group of 20 cases treated institutionally revealed the following changes during the course of treatment (Macrae, 1947).

(i) Accentuation of the congestive aspect of the disease during the first 3 weeks and, in a few cases, the development of spontaneous ulcers ; (ii) increase in the diameter of the Mantoux reaction to 1 in 1,000 and 1 in 10,000 tuberculin during this phase, with subsequent return to the original, or to slightly less than the original diameter; (iii) increase in the sedimentation rate during the first 2 months, followed by a gradual return to normal, and (iv) a slight polymorphonuclear leucocytosis occurring about the same period.

Early aggravation of symptoms

Tuberculin reaction during treatment with calciferol

Macrae (1947) found that local therapy after a period of treatment with calciferol alone greatly hastened progress. Toxic symptoms developed in half the cases. Of the first 20 cases, 14 were discharged within 6 months, clear of all active disease.

Toxic effects of calciferol

Toxic symptoms which may be met with during the course of treatment with calciferol in heavy dosage are nausea, epigastric discomfort, depression, weakness, polydipsia, polyuria and constipation; the last is particularly common in children. These symptoms may be serious, and the first signs call for either a temporary cessation of treatment or a reduction in dosage. Their interpretation is not yet clear. Ingram and others (1946) have stated that they are always associated with a rise in the diffusible calcium content of the blood-serum. Conversely, high total serum-calcium figures are met with quite often in the course of routine estimations, without symptoms of toxicity. One case (Bureau, 1945) of deposition of calcium in and about finger joints

Toxic effects

Serum-calcium figures

has been recorded. Obviously, careful supervision is required during treatment with the massive doses of calciferol needed for the treatment of lupus Serum-calcium estimations in particular should be made at regular intervals; treatment should be stopped temporarily when the figure has risen to 12 milligrams per 100 cubic centimetres or higher and should not be reinstituted until a normal reading is obtained (9·5–10 5 milligrams per 100 cubic centimetres), usually in from 1 to 3 months.

8. HISTOLOGICAL CHANGES IN LUPUS TREATED WITH CALCIFEROL

These changes consist essentially of the development of a quantity of young connective tissue in and about the lupus nodules. This delicate-looking fibrillary tissue seems to fragment the process, and ultimately to replace it. The

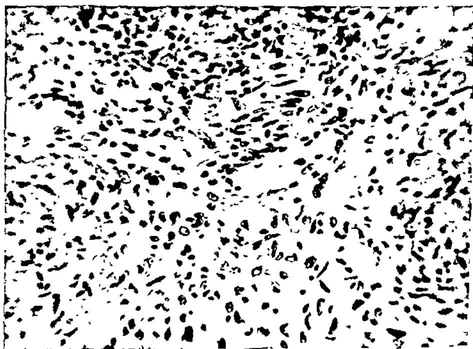


Fig. 214 —The same tubercle as in Fig 213, showing a few giant cells and endothelial cells (× 420). (*Brit. med. J*)

giant-cell systems (Fig. 214) are first invaded by histiocytes; these cells are seen between the endothelial cells (Fig. 215). This is followed by zones of early fibrous tissue which appear in and about the giant-cell systems, isolating them from one another (Fig. 216). Somewhat later the process of disintegration of these systems has advanced and individual giant cells may be seen separated from one another by new connective tissue. Finally, the endothelial cells and giant cells disappear and are replaced by fibrous tissue (Figs. 217 and 218).

9. LOCAL TREATMENT

Somewhat less than half of the cases of lupus may be expected to clear up under vitamin D therapy without the help of local treatment. For the

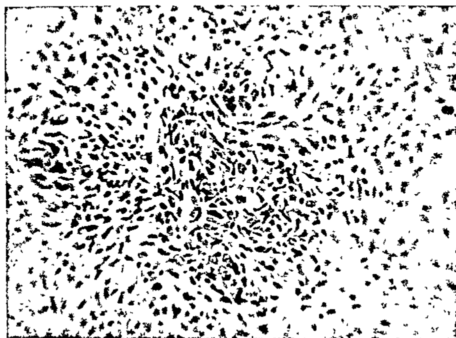


FIG 215.—Same case as in Fig 213, 16th April Invasion of the tubercle by histiocytes (× 420). (*Brit. med. J.*)



FIG. 216.—Same case as in Fig. 213, 30th April Development of fibrous tissue separating the giant-cell systems from one another (× 250). (*Brit med J.*)



Fig. 217.—Same case as in Fig. 213, 21st May. Separation of giant cells by the newly formed connective tissue (× 420). (*Brit. med. J.*)



Fig. 218.—Same case as in Fig. 213; also 21st May. Showing separation of tubercles from each other by connective tissue producing an appearance similar to that seen in sarcoid; also disappearance of endothelial cells from the tuberculous lesions (× 90). (*Brit med. J.*)

remainder, local measures are necessary. These, however, need not be instituted until the disease has been brought well under control. The following may then be adopted

(1) **Finsen therapy**

In the Finsen lamp or one of its modifications, light from a 20-ampere carbon-arc lamp is focused through a series of telescopic lenses made of quartz on to a water-cooled quartz compressor, which is held in position on the small area of skin to be treated. A skilled technician is required for this treatment which varies in duration from 30 minutes for disease near the eye in a sensitive patient, up to as much as 2 hours for other regions. A reaction which persists for 2 or 3 weeks is produced. Light so applied has a selectively destructive action on lupus nodules. The cosmetic result is excellent.

Cosmetic result

(2) **Kromayer treatment**

The Kromayer lamp is a water-cooled mercury-vapour lamp with various solid quartz applicators and is of considerable value in producing a superficial reaction in a short time, that is, in ½–5 minutes according to individual sensitivity of the skin to this type of irradiation. It may be used to initiate a reaction in tough scarred areas which afterwards will be treated by the application of a selective caustic.

(3) **Selective caustics**

Of these the little-known picric brass paste is one of the most satisfactory. It is an unstable preparation consisting of 85 per cent basic copper sulphate, 14 per cent zinc sulphate and 1 per cent picric acid. It is applied under strapping and removed after 24 or 48 hours. It has the effect of eating into the nodule as it oxidizes, leaving the normal skin undamaged. Several applications may be necessary. Liquid acid nitrate of mercury may be painted over small areas and afterwards neutralized, for example, with soft soap, or it may be pricked into lupus nodules with a pointed match-stick. Several nodules may be treated at one time. After 24 hours this treatment may be completed by an application of Kromayer light and brass paste.

Method of application

(4) **Electrocoagulation**

Lupus inside the nose, mouth and pharynx may be treated by diathermy. This type of local treatment is commonly used also for resistant lupomas of the skin, particularly in France.

(5) **Curettage of vegetations**

Fungating lesions are relatively resistant to treatment with vitamin D and are best dealt with by scraping thoroughly under general anaesthesia. The resulting raw area is swabbed with pure lysol to prevent bleeding and to promote an inflammatory reaction; alternatively, potassium permanganate crystals may be applied and the resulting scab allowed to separate spontaneously.

General anaesthesia

The raw area left after any of the above local measures may be dressed with a sedative ointment such as Treloar's paste, having the following formula:

Zinc oxide – –	10 parts
Vaseline – –	40 parts
Lanolin – –	50 parts

10. INDICATIONS FOR SURGICAL INTERVENTION

Repair of mutilation

The surgical treatment of lupus is concerned chiefly with the repair o destructive inroads of the disease on important tissues—for example, g narrowing of the mouth or nostrils, destruction of the nose, blocking o lacrimal channel and ectropion. In general, plastic surgery is contra-indic unless the patient is cured, for recurrence of lupus in the grafted skin example, of a reconstructed nose, has proved to be extremely resistar treatment. In the case of ectropion, however, if the eyelids fail to meet du sleep, it is advisable to operate immediately in order to avoid the occurr of keratitis and possible loss of sight. It may also be desirable to co microstomia early in order to facilitate feeding or dental treatment.

The surgical measures usually adopted are as follows:

(1) For ectropion, Thiersch grafts from the inner side of the upper arm applied over Stent moulds.

Free grafts

(2) For deformities caused by fibrosis, Thiersch grafts may be used to c raw areas after removal of the scarred and contracted skin.

(3) For lupus complicated by radiodermatitis, excision of the diseased is followed by a Thiersch graft.

(4) The correction of deformity caused by active destruction of a part. is most often needed in the case of the nose. Rhinoplasty is usually car out by turning down a frontal flap to construct a new nose, with or witl cartilage to give support, the raw area on the forehead being filled in l Thiersch graft. Alternatively, a tube pedicle from the abdomen can employed.

Rhinoplasty

Epiphora

(5) *For epiphora. Often a mild degree of ectropion will pull the lacri* opening away from contact with the eyeball and scarring may block the na lacrimal duct. Probing of the lacrimal apparatus with enlargement inward the punctum is often effective. A cocoon graft, using mucous membr from the mouth, may be required to replace the conjunctiva.

(The microphotographs in Figs. 213–218 were taken by Mr. A. E. Clark, technician to Department of Pathology at St. Thomas's Hospital.)

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[References to other titles are given under Lupus Vulgaris, in the Index Volu The subject is also dealt with in the *British Encyclopaedia of Medical Prac* (1938), Vol. 8, p. 254.]

LYMPHOGRANULOMA INGUINALE

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1. DEFINITION

227.] Lymphogranuloma inguinale is a virus disease of venereal origin occurring predominantly amongst Negro races in the tropics. It is associated with general constitutional disturbance of varying degree and with local manifestations which differ in the male and female.

2. NOMENCLATURE

Though lymphogranuloma inguinale is the name now most commonly used, the disease is variously known in different parts of the world as "tropical bubo", "climatic bubo", "lymphogranuloma venereum", "poradenitis" and by other names. This multiplicity of terms has led to much confusion, since, in the past, each was thought to be either a separate entity or some bizarre manifestation of syphilis. The introduction by Frei in 1923 of the intradermal antigen test (Frei, 1925) did much to clarify the position and, indeed, to show that yet other lesions, such as "*esthiomène*" and "proctitis leading to rectal stricture", were of the same origin.

3. AETIOLOGY

Racial incidence

Lymphogranuloma inguinale has been called by Stannus (1933) "a sixth venereal disease". It affects the Negro races in particular and especially in the tropics, but it is encountered in temperate climates wherever large communities of Negroes live, as for instance in the United States of America and in South Africa; rarely, it occurs in white people. Its incidence varies as the sexual promiscuity of the population, and where prostitution and polyandry are rife it is no uncommon thing to find natives suffering from two or even three venereal diseases at the same time. The disease occurs at the height of sexual activity, men being affected more commonly than women. Recently it has become more widespread outside the tropics, and Armstrong and Niebauer (1944) quote an incidence of 10 8 per 10,000 admissions to the New York Hospital in 1940. Although almost always of venereal origin it occasionally results from close contact, as in children sleeping in the same bed as an infected adult, or from direct infection, as when a surgeon or laboratory worker pricks his finger when dealing with tissue from a case of the disease.

Age incidence

Mode of infection

4. SURGICAL ANATOMY

Since the characteristic features of the disease are largely due to glandular involvement, the anatomy of the lymphatic drainage of the genitalia is important. In the male the primary lesion is on some part of the external genitalia, the lymph drainage from which is the inner end of the inguinal group of glands. In women two sets of lymphatics may be involved. If the primary lesion is on the clitoris or vulva the glands affected are again the inner ones of the inguinal group, but if, as is much more common, the primary lesion is on the vaginal wall or cervix, the pararectal and retroperitoneal glands are the ones infected, and this leads to the so-called genito-ano-rectal syndrome. As a result of homosexual practices this syndrome occasionally affects men.

In the male

In the female

5. PATHOLOGY, MORBID ANATOMY, BACTERIOLOGY

(1) The virus

In 1913 Durand, Nicolas and Favre recognized lymphogranuloma inguinale as a separate entity. In 1923 Frei demonstrated a specific antigen, and in 1930 Hellerström and Wassen (1931) discovered a filtrable virus which could be transmitted to monkeys and mice, and which was not unlike that of psittacosis. Some workers claim to have seen inclusion bodies, but others deny this.

(2) The primary lesion

As is mentioned above, the primary lesion in both men and women is on some part of the genitalia, and it is of fleeting character, leaving little or no residual scarring. It takes the form of tiny vesicles, papules or ulcers of a herpetiform nature developing within 24–48 hours of sexual intercourse.

(3) Secondary lesions

The characteristic feature of the disease in men is the glandular mass in the groin and in women the genito-ano-rectal syndrome.

(a) The glands in males

About 3–8 weeks after infection a gland in one or both groins enlarges, usually at the inner end, though not uncommonly at the middle and rarely at the outer end. At first it is solitary and non-adherent to the skin or deeper structures, but later on surrounding glands become involved, which, together with periadenitis, form a hard brawny mass. If sectioned at this stage the glands present a very characteristic appearance in that they look as though packed with tiny abscesses. A little later the central part of the mass softens in several places, the result of coalescence of these abscesses, and on section the softened areas consist of cavities broken up by interlacing trabeculations of fibrous tissue, and containing sterile glairy fluid which sticks to the knife. Finally the mass breaks down spontaneously leaving discharging sinuses. Microscopically, the appearance is that of granulomatous tissue with giant-cell formation and areas of necrosis, and it is easily confused with a tuberculous process.

The bubo

Breaking down

(b) Genito-ano-rectal syndrome in females

The vulva becomes oedematous and surmounted by condylomatous masses, from cracks and fissures within which exudes an evil-smelling discharge. This is the condition called *esthiomène*. Similar masses may arise from the anal region and, as the perirectal lymphatics become infected, infiltration and fibrosis of the rectal wall leads to the characteristic stricture. Typically, this stricture is about 1 inch from the anal margin and it extends upwards for a further 2 inches in a funnel-shaped fashion. At first the mucous membrane is congested and oedematous, but later on it becomes grey, rigid and ulcerated. Microscopically, the mucous membrane and muscle are replaced by fibrous tissue containing polymorphs, eosinophils, mast-cells and plasma-cells, and Wright, Freeman and Bolden (1946) state that there is a palisade formation of epithelioid cells which is pathognomonic.

Esthiomene

Stricture

(4) Frei's test

The recognition by Frei of a specific antigen was a discovery of the greatest importance He aspirated a softening gland and treated the material by heating to 60° C. for two hours on one day and then again for one hour on the following day. This material was then diluted with saline and the patient to be tested was given an intracutaneous injection of 1–2 minims of the mixture. A red raised flare at the site of injection, developing within 24–48 hours, was regarded as a positive result.

6. CLINICAL PICTURE

(1) The primary lesion

The symptoms commence within 24–48 hours of intercourse, with small painless papules, vesicles, pustules or ulcers on some part of the genitalia.

In the male the usual site is on the glans, corona or undersurface of the prepuce, but it may be on the shaft of the penis or even on the scrotum. The rate of incidence is not any lower in the circumcised. It has been stated that a nodular condition of the lining of the distal urethra develops, but this was not revealed by urethroscopy in a series of cases investigated by myself and my colleagues when serving in Sierra Leone during World War II.

In the male

In the female

In the female the commonest site for the primary lesion is on the cervix and walls of the vagina, but it may also affect the clitoris or labia.

Short duration

In either sex the primary lesion is of a fleeting nature and, since the essential features of the disease do not develop until 3–8 weeks later, the patient may

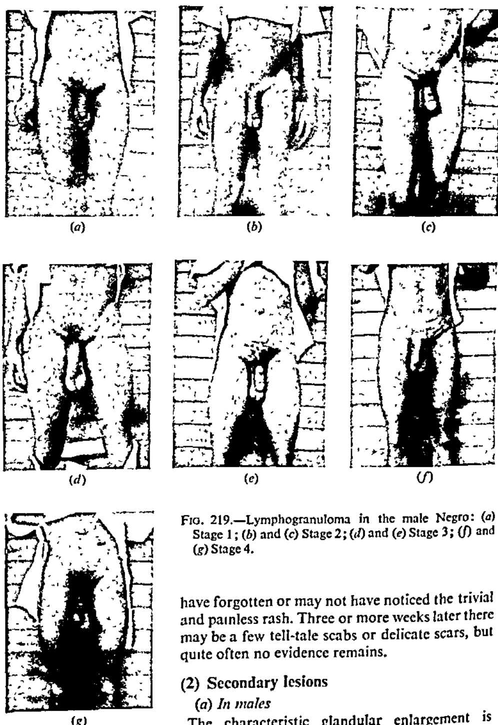


FIG. 219.—Lymphogranuloma in the male Negro: (*a*) Stage 1; (*b*) and (*c*) Stage 2; (*d*) and (*e*) Stage 3; (*f*) and (*g*) Stage 4.

have forgotten or may not have noticed the trivial and painless rash. Three or more weeks later there may be a few tell-tale scabs or delicate scars, but quite often no evidence remains.

(2) Secondary lesions

(*a*) *In males*

The characteristic glandular enlargement is described above under "Pathology". The following classification, introduced by Law and myself (Stammers and Law, 1942) as the result of an experience of over 150 cases, and confirmed by me

(Stammers, 1943) after a further experience of up to 300 cases, is useful in deciding prognosis and treatment.

Stage 1.—A solitary firm elastic gland is observed, free from adhesions to skin or deeper structures, which is rarely tender, though physical activity may cause pain. (Fig. 219 (a).)

Stage 2.—The gland becomes adherent to skin and deeper structures, and surrounding glands begin to enlarge and become fused with the original one. This develops into a hard brawny painful mass, covered by dusky red skin showing *peau d'orange*, but the bubo does not fluctuate at this stage. The constitutional disturbance may be considerable, and it is always greater in white people, in whom external and common iliac glands may also be enlarged, though I have never seen these break down. Malaise, pulse of 100

The bubo

Constitutional disturbance

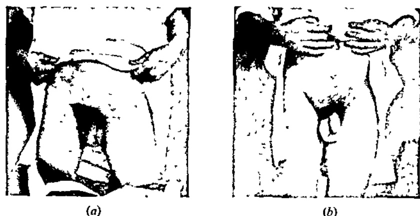


FIG 220.—Lymphogranuloma in the male European (*a*) Stage 2, (*b*) Stage 3.

per minute and temperature up to 103° F. are common, and anaemia, bronchitis and jaundice may develop, the two last-mentioned possibly resulting from pressure by enlarged glands. (Fig. 219 (b) and (c) and Fig. 220 (a))

Stage 3.—The original mass softens in one or more places and the general condition improves. (Fig. 219 (d) and (e) and Fig. 220 (b).)

Stage 4.—Sinuses develop, and secondary infection leads to discharge, which lasts for weeks or months on end, severe anaemia being a common accompaniment. (Fig 219 (f) and (g).)

It is easy to mistake the swelling for tuberculous lymphadenitis, especially when cough, anaemia and wasting are present.

(*b*) *In females*

The condition of *esthiomene*, with its noisome discharge, is most distressing, and the constitutional upset may be considerable. The characteristic feature, however, is rectal stricture, following which, apart from symptoms of chronic intestinal obstruction, fistulae develop between rectum and vagina, rectum and ischio-rectal fossa and even rectum and bladder, each complication producing its own particular features. On digital examination of a strictured rectum the finger feels as if it were inside an onion from which the heart had been removed—the rectum is both rigid and conical.

Esthiomène

Stricture of rectum and fistulae

7. SPECIAL AIDS TO DIAGNOSIS

Frei's antigen test is described on p. 489.

8. DIFFERENTIAL DIAGNOSIS

Other infections

Regarding enlarged glands, it is, of course, essential to exclude any other source of infection, especially because tropical ulcers and skin yaws are so common in natives; a chafing boot or patches of epidermophytosis are common in white races. All other venereal diseases—gonorrhoea, syphilis, chancroid and yaws—must be considered, as also tuberculous or other types of more generalized adenopathy. In stage 4, actinomycosis has to be excluded.

Positive Frei test

In brief, it may be said that in the absence of any other source of infection a mass confined to one or both groins, conforming to any of the four stages described above and with a positive Frei reaction, means lymphogranuloma inguinale. The test will also differentiate lymphogranuloma inguinale in the case of lesions of the genito-ano-rectal type.

9. PROGNOSIS

Elephantiasis

Stricture

With appropriate treatment most of the lesions will clear up leaving little more than a residual thickening. The reaction to the Frei test, however, tends to remain positive for many years. Vulval lesions may lead to elephantiasis in spite of adequate treatment, and rectal stricture and its fistulous complications become problems requiring surgical relief.

10. TREATMENT

(1) Males

There is no doubt that, in natives with typical bubo, many cases never pass beyond stage 1 and subside spontaneously without the victim's ever "reporting sick". It is also a fact that in other cases in stage 1 in which the patient "reports sick", the disease clears up with a few days' rest. The difficult cases are those in stage 2, since the patients tend to linger on in this condition for many weeks during which there is considerable general upset. Active treatment leads either to rapid subsidence or rapid progression to stage 3. In white races there is little chance of even stage 1 cases clearing up spontaneously.

(a) *Medical treatment*

The following are the lines of treatment available:

(i) *Rest in bed with local applications of heat* in the form of fomentations, Antiphlogistine, radiant heat or infra-red heat.

(ii) *Intramuscular injections of Anthiomaline* (10–20 injections) as described by Law (1942). This preparation is a solution of lithium antimony-thiomalate and is given in doses of 0·5 millilitre increasing up to 2 millilitres (1 millilitre = 0·01 gramme Sb). It is a very satisfactory treatment and its great advantage is that patients can be treated as ambulatory cases.

(iii) *Intravenous injection of sodium antimony tartrate.*—This is effective but painful and may lead to sharp reactions; it therefore has to be given as in-patient treatment.

(iv) *Large doses of sulphonamides.*—As much as 96 grammes, spaced over 3–4 weeks, in 3 courses, was used in Sierra Leone during World War II, both sulphanilamide and sulphapyridine being effective. Intramuscular injections of a suspension of sulphapyridine, using about one-quarter of the above-mentioned dosage, were also found satisfactory, but a watch has to be kept on the leucocyte count, and it is important to avoid injections in the vicinity of the sciatic or other main nerve.

(v) *Artificial pyrexia,* as produced by the intravenous injection of a 50-million dose of T.A.B. vaccine, has proved to be of value in a certain number of cases but is quite ineffective in others.

(vi) *Penicillin.*—No large series of cases has been treated by penicillin in West Africa, and I am indebted to members of the Colonial Medical Service for this information. On general principles one would expect it to be useful in combating secondary infection, but unlikely to have a specific effect on the virus.

(b) Surgical treatment

The only type of case in which excision is likely to be successful is that in stage 1. These lesions heal well and recurrence is unlikely. Early stage 2 cases may be successful, but a number of them break down, and later cases invariably break down. In stage 3 there is a natural temptation to incise the fluctuating mass, but this always leads to secondary infection and a protracted illness, and it should be avoided at all costs The best treatment for this stage is aspiration, which may have to be repeated, combined with Anthiomaline or a sulphonamide, though, curiously enough, the use of a seton will bring about cure, the method being popular among medical missionaries in China.

(2) Females

For lesions in females, chemotherapy is again the best treatment, though, as already explained, the genito-ano-rectal syndrome may necessitate colostomy in order to relieve stricture or the effects of fistulation.

11. RESULTS OF TREATMENT

A small proportion of bubos recur and require a second course of treatment. In females the potentialities are far more serious and carry all the risks and disappointments common to such complications.

(I wish to thank Mr. W. A. Law for his kind permission to use the photographs in Figs. 219 and 220, which are those used in his thesis for the Mastership in Surgery of the University of Cambridge, 1942.)

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[References to other titles are given under Lymphogranuloma Inguinale in the Index Volume. The subject is also dealt with under the heading of Lymphopathia Venereum in the *British Encyclopaedia of Medical Practice* (1938), Vol. 7, p. 287.]

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